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Fortis

Clinical Connect

Fostering a culture of innovation and excellence

Paediatrics and Neonatology Special

It Takes a Lot to Treat the Little Ones

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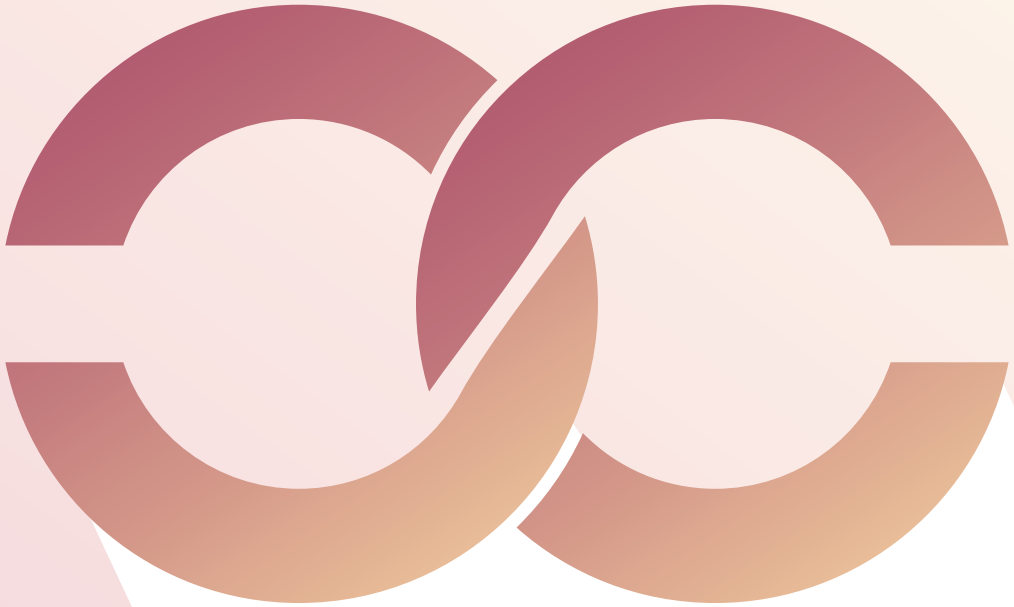

INSPIRATION MESSAGES

Dr Narottam Puri
Advisor (Medical),
Fortis Healthcare Limited

This edition of "Clinical Connect" is dedicated to Paediatric Sciences. The word, "Paediatrics" is the art and science of treatment of children's ailments and the word is of Greek origin - Pais meaning 'child' and iatric meaning a healer (doctor).

The branch of Paediatrics emerged from Medicine (Internal Medicine) when it became obvious that Paediatrics patients had unique needs, they behaved and responded differently to medical treatment, and required a greater degree of care and caution than adults.

In India, George Coelho in 1928 made a beginning by practicing and propagating Paediatrics as the Superintendent of BJ Hospital for Children - the first children's hospital in India and is considered the Father of Indian Paediatrics. In USA this honor belongs to Abraham Jacobi whose work, teaching and writing in the latter part of 19th Century made him the Father of American Paediatrics.

The frontier of paediatrics have further expanded through development of subspecialities - Paediatric Cardiology, Paediatric Neurology and, Paediatric Gastroenterology through Allergy and Immunology Infectious Disease and all other adult subspecialities. Lately, lot of attention has focused on Paediatric Haematology, Oncology and Bone Marrow Transplantation.

At Fortis, we have the right experts with adequate experience and acumen to handle all types of Paediatric and Neonatal patients with some centres emerging as Centres of Excellence in the field.

Statistics indicate that in 2021 approximately 26% of India's population is in the age group of 0-14 years and 31-35% Indian population is in 0-18 years bracket. A specialty that caters to about 1/3rd of the population of the country is of critical importance and I am delighted that Fortis is playing a vital role in the care of our children.

This issue of Clinical connect is focusing on the future of our country and highlights the work and experience of many of our colleagues at Fortis.

I take this opportunity to congratulate FMC, Fortis clinical fraternity, MSOG and the editorial board for this 10th issue of Clinical Connect. It is testimony to the dedication, commitment and ownership towards this unique Fortis Initiative.

Clinical Connect has become beacon of Clinical Talent, providing a platform to our clinicians to showcase their clinical and research work and to share best practices to benefit patients and caregivers.

Dr Krishan Chugh
Principal Director and Head - Paediatrics and PICU,
Fortis Memorial Research Institute, Gurugram

Many surveys have concluded that among clinicians, Paediatricians have the highest happiness and professional satisfaction quotient. Likely explanations include better outcomes in childhood illnesses, the innocence of their clients (children), and the enthusiasm of the young parents. And most importantly, a good part of their work is with healthy children- immunization, growth monitoring, etc. Some believe that Paediatrics as a specialty is chosen by only those who have a softer personality. Whatever the reasons I have enjoyed my long innings and I continue to do so. This is especially because I have spent more than three decades in Paediatric Critical Care. Complete recovery rates are much higher in critically ill children than in adults.

Sub-specialties of Paediatrics have evolved comparatively recently and have certainly resulted in a better understanding of childhood illnesses by the respective specialists. We are fortunate to have well-developed sub-specialties in several of our hospitals in the Fortis network, adequately supported by appropriate equipment and technology. At Fortis Memorial Research Institute, Gurgaon we are able to provide the highest level of services to the sickest of the children referred to us from various parts of our country as well as from abroad. Liver and kidney transplants are done regularly with rewarding outcomes. Bone marrow transplant numbers and success rates compare very well with the best in the world.

Paediatric Pulmonology services at Fortis are of the highest class in the country. As we provide diagnostic as well as therapeutic procedures for preterm, infants, children, and adolescents supported by appropriate, equipment and skills. Balloon dilatation of stenotic lesions of airways, cryoablation, cryo-biopsies of the lung, and glue therapy, are some of the unique services provided here. Our Centre has one of the largest experiences in removing foreign bodies from the airways of young children, using flexible bronchoscopes under sedation.

Teaching and training are integral part of any advanced medical centre. Many of our centres in the Fortis network are providing DNB degrees in general paediatrics and fellowship programs in some of the sub-specialties of paediatrics like paediatric critical care. Efforts are also being made to provide certified in-job training to nurses in paediatrics, neonatology, and other specialist units of paediatrics. We also take pride in being at the forefront of providing continuing medical education programs for the general paediatricians in our region and conducting workshops for fellow trainees in paediatric critical care and neonatology.

"Radiation Safety in Paediatric Imaging - safe and effective imaging care of Children"

Dr Arvind Kumar
Chairman - Fortis Paediatric Speciality Council
Director and Head - Paediatrics,
Fortis Hospital, Shalimar Bagh, New Delhi

Dear Colleagues,

I congratulate the dynamic editorial team of clinical connect for opening the frontiers of Paediatrics and neonatology in the current issue.

Paediatrics is a branch which enables us to touch the lives of hundreds of families. We have the singular opportunity where we can say that as a paediatrician, we feel pride in nurturing the future of humanity.

Modern Paediatrics as a branch in India has its origin in Mumbai in 1928 under the leadership of George Coelho, called the father of Paediatrics. Since that time Paediatrics has branched into different subspecialties and I am proud has branched into different subspecialties and I am proud to say that in Fortis, these specialized sections are well developed and offer new therapies and diagnostics for the welfare of our little patients.

Each step in our field will bring new expectations and unavoidable challenges in these rapidly evolving times which must be met collectively and constructively. Challenges waiting for a tangible solution are:

- Changing behaviour of pathogens ensuing changing disease patterns
- Emerging drug resistance
- Increasing incidence of psychological and developmental problems in children and adolescents
- Disturbingly growing lifestyle diseases - obesity, type 1 diabetes, and hypertension
- Impact of environment on child health
- Upgrading of technology, with tremendous potential of AI in the field of healthcare

The current issue of Clinical Connect will be extremely beneficial to the reader and will provide insight into the work and research of our colleagues at Fortis. Wishing you all good health and an optimistic future ahead.

Dr Raghuram Mallaiah
Senior Director - Neonatology,
Fortis La Femme, GK II, New Delhi

In the rapidly evolving landscape of healthcare, collaboration among professionals across various disciplines has emerged as a vital catalyst for driving medical progress. Fortis Clinical connect provides an ideal platform for healthcare professionals to share their expertise, knowledge, and innovation to bring about transformation in patient care, improve interdisciplinary collaboration and advance research and innovation, all of which are the backbone of a robust healthcare system.

Fortis Clinical Connect, by bringing together experts from diverse backgrounds, including physicians, nurses, researchers, and administrators, will help in harnessing collective intelligence to improve patient outcomes. Furthermore, continued professional development programs such as this can empower healthcare providers to stay abreast of the latest advancements and best practices in their respective fields.

By actively encouraging the submission of interdisciplinary research papers, case studies, and commentaries, Fortis Clinical Connect can act as a powerful catalyst to facilitate the dissemination of knowledge across disciplines and help us clinicians to keep abreast of the latest advancements and best practices in our respective fields.

Lastly Fortis Clinical Connect can act as platform for connecting researchers, clinicians, and other professionals, enabling networking, and facilitating collaborations not only within the Fortis network but even on a global scale.

Dr Jesal Sheth
Consultant - Paediatrics,
Fortis Hospital, Mulund

Over the years, Paediatrics at Fortis has evolved into a robust department, we are specialized in handling complex cases and consistently delivering excellent outcomes, particularly in various surgical Paediatric specialties. Our success can be attributed to several factors, including our multidisciplinary approach, accessibility, expertise, and abundance of resources. As a Senior Paediatrician at Fortis Hospital, Mumbai I am honoured to share some remarkable achievements and the values we uphold in our Paediatric department.

Working with Fortis healthcare is an extraordinary privilege. There is a synergy between our team of expert clinicians and hardworking nurses, empowered by a dedicated administration which creates an environment of endless possibilities. Here, we engage in open conversations, tackling challenges head-on, constantly learning and unlearning through comprehensive training programs. We forge a unified path, committed to our common goals. Our dedication goes beyond mere job performance; we extend our support to families, actively involving them in the care of sick neonates and children. This compassion, coupled with our unwavering pursuit of medical excellence, has yielded remarkable clinical outcomes.

Even during the ongoing pandemic, we stood out as the only private hospital equipped to handle paediatric COVID-19 and MIS-C cases. Our team successfully managed numerous critically ill children and neonates, showcasing our resilience and unwavering dedication to providing exceptional care in challenging circumstances.

Clinical Connect is a great platform, as we have an additional avenue to share our diagnostic and therapeutic challenges with the wider Fortis family. I encourage you all to embrace this opportunity and participate actively in sharing your research work and clinical contributions to the organisation. These endeavours aim to foster knowledge sharing, exchange experiences, and leverage our strengths, ultimately facilitating continuous learning and growth.

I extend my heartfelt wishes to all of you as you embark on this remarkable initiative. May it lead to even greater achievements, as we continue to deliver exceptional care with transparency and compassion.

DA VINCI XI ROBOTIC SYSTEM
Date: May 03, 2023
Location: Fortis Hospital, Noida

Dr Mala Airun
Director - Medical Operations,
Fortis Escorts Hospital, Jaipur, Kolkata and Chennai

I take this opportunity to thank the MSOG team and our group of clinicians for this initiative. This edition of Clinical Connect fantastically puts together all the interesting case studies and interesting clinical work accomplished during the past 1 year in Paediatrics Sciences.

We at Fortis are guided by the purpose to provide special care for expecting mothers and our paediatric population. Our **"Paediatrics Specialty Council" comprises an inspiring group of clinicians who come together to make this possible.**

This has been an impactful year for the paediatric specialty council. We worked on a vaccination and hand hygiene awareness drive in schools and finalized Paediatric early warning signs for the nurses (to be rolled out by July). We have seen some new programs, as well as the growth of the specialty.

-Neonatal Resuscitation Program for nurses where all the select nurses assigned to the Labour room, OT, NICU, PICU, SICU, and ER were trained in resuscitation techniques for neonates and paediatric patients according to the IAP guidelines at the Anandpur and Jaipur units.

-Adolescent Health Program at schools based on Fortis School Mental Health Programs where sessions on Bully to Buddy, Panchatantra tales and problem-solving, Stress management for teachers, Good touch and Bad touch, and Study and Exam skills were conducted by the Department of Clinical Psychology

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Please send your comments, feedback and suggestions to
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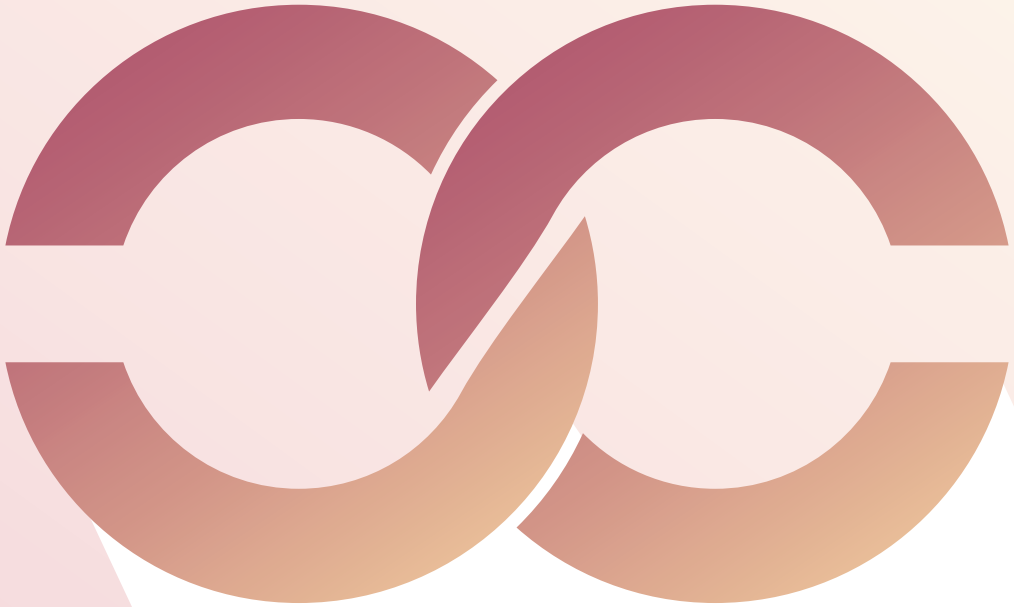

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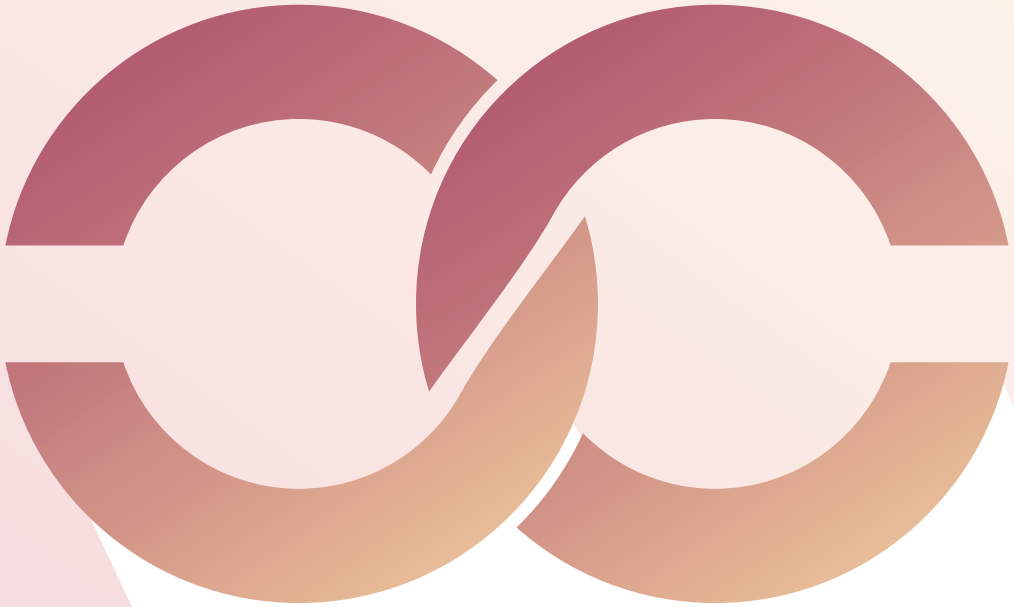


LEADING THE WAY AT FORTIS

Liver Tumour Diagnosed Antenatally in a Baby

Dr Vikas Dua
Director and Head - Paediatric Hematology, Fortis Memorial Research Institute, Gurugram

Dr Anand Sinha
Consultant - Paediatrics, Fortis Memorial Research Institute, Gurugram

Introduction

Diagnosis and treatment of neonatal liver tumors is challenging. This case represents a successful approach for the management of a rare tumor called as congenital Hepatoblastoma. The rarity of this tumor warrants this report. The case also illustrates a personalized treatment approach and multidisciplinary care resulting in an excellent outcome.

Case Description

Baby Kshirsa Rimal Shrestha, resident of Nepal, presented to us at Fortis Memorial Research Institute, Gurugram on day 8 of life with history of a solid mass in liver which was picked up incidentally on an antenatal scan. USG done at birth showed a solid mass measuring 4.2x 2.8 x 2.8 cm in segment VI and VII of liver displacing right hepatic vein. MRI abdomen was done which showed 4.3x 2.8x 3.6 cm mass in segment VI and VII of liver abutting right hepatic vein and inferior vena cava consistent with Hepatoblastoma (pretext I). Serum AFP level was high (125731 ng/ml). She underwent USG guided biopsy here which was suggestive of pure fetal type Hepatoblastoma. Whole body PET CT scan was done which was suggestive of localized disease with no distant metastasis or vascular invasion.

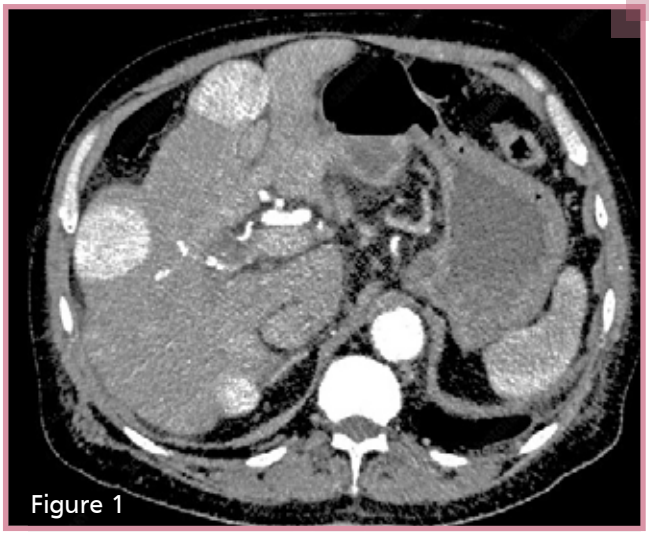


Figure 1

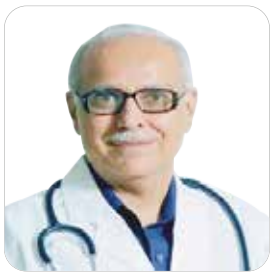
Treatment

She started on cisplatin-based chemotherapy. Serial AFP monitoring was done along with chemotherapy which showed progressive decline (AFP was 13316 ng/ml after 1st cycle and 9211 ng/ml (04.07.2022). Repeat scan after 4 cycles showed decrease in size of the lesion. After 4 cycles of chemotherapy she underwent surgical resection by Dr Anand Sinha (paediatric surgeon). Post-surgery she received two more cycles of adjuvant chemotherapy. To avoid chemotherapy induced toxicity the dose of cisplatin was adjusted as per her weight. During chemotherapy, the patient did not experience any major side effects and tolerated treatment well.

The prognosis is excellent for children with standard risk hepatoblastoma who have a good response to chemotherapy and good resection. Presently the patient is 11 months old and growing well. She is under follow-up of paediatric oncology services.

Figure 2

Primary Ciliary Dyskinesia and Situs Inversus Totalis in a Neonate- A Case Report



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Introduction

Primary ciliary dyskinesia is an autosomal recessive ciliopathy characterized by chronic sinopulmonary infections, asthenospermia, and immotile cilia. Respiratory epithelial cells and sperm flagella of affected individuals lack both the inner and outer dynein arms. About 50% of patients have situs inversus.

We report a neonate with respiratory distress who was diagnosed with situs inversus totalis during admission and results of NGS, next-generation sequencing; confirmed Primary Ciliary dyskinesia.

Case Report

A male baby was born by LSCS at 38 weeks 4 days at a nursing home. Baby cried immediately after birth and required no active resuscitation. Birth weight was 2800 grams. At about 48 hours of age, baby developed respiratory distress, hence referred to Fortis Hospital. On admission, baby had respiratory distress. SpO2 on room air was 79%, which increased to 92% on Oxygen by hood at 10 litre/min. RBS at the time of admission was 220mg/dl. Baby was immediately admitted to NICU for further treatment and management. During the stay child had severe respiratory distress and required heated humidified high flow nasal cannula (HHHFNC) support.

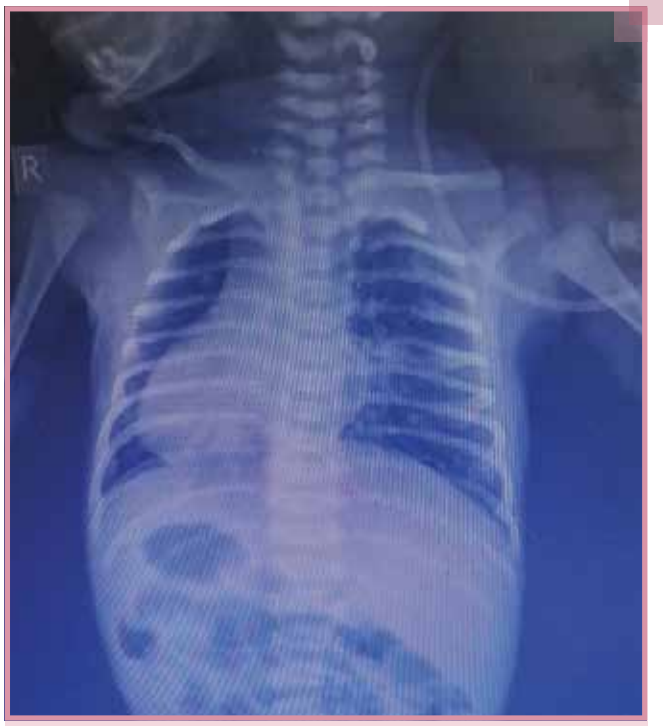

Figure 1 : Chest x-ray showed dextrocardia with situs inversus with B/L heterogenous opacities

HHHFNC support was given for eight days. Baby had respiratory distress/ tachypnea which gradually settled over periods of 12 days. Oxygen support was given by Head box / Nasal prong for three days thereafter. After that baby was on spontaneous breathing on room air.

During stay, shifting lung collapse was noted (one day chest x-ray showed left upper lobe collapse and next day showing right upper lobe collapse)

Subsequent chest x-rays indicated situs inversus with mediastinal shift. To address these findings, the medical team initiated chest physiotherapy and nebulization with normal saline. The baby did not exhibit any cardiovascular issues, bradycardia, or desaturation. A 2D-Echo was performed, revealing dextrocardia with situs inversus, a right arch, and a patent foramen ovale with left-to-right shunt , PFO(L□R), but no other cardiac anomalies were detected.

Baby had no neurological concerns throughout stay. Inj Taxim and Inj Amikacin were started for initial five days in view of presumed sepsis. Partial septic screen was sent which was normal. Septic screen was repeated which was negative following which antibiotics were stopped. Blood culture showed no growth after five days. Initial

one day child was on IV Fluids. Orogastric feed was started then and continued for nine days and after that breast feed was initiated. The child was in the hospital for two weeks and was discharged when general condition became stable and was maintaining saturation on room air. The baby exhibited good crying and activity levels and demonstrated acceptance of and tolerance for breastfeeding at the time of discharge. Based on geneticist opinion whole exome sequencing (WES) was recommended and results were obtained after 5-6 weeks.

Results of WES showed homozygous single base pair deletion in exon 5 of the LRRC6 gene (chr8:g.132632763del; Depth: 102x) that results in a frameshift and premature truncation of the protein 12 amino acids downstream to codon 210 (p.Trp210CysfsTer12; ENST00000620350.5) was detected . The observed variation has previously been reported in patients affected with ciliary dyskinesia. The variant has not been reported in the 1000 genomes database and has a minor allele frequency of 0.0006%, 0.06% in the gnomAD and our internal databases respectively. The reference region is conserved across species.

Follow Up

Child is now 14 months old and his physical and mental developments are normal. No history of sinopulmonary infections, otitis media or hospitalization.

Discussion

Primary ciliary dyskinesia manifests clinically as chronic bronchitis leading to bronchiectasis, chronic rhino-sinusitis, chronic otitis media, situs inversus (in approximately 50% of cases), and male infertility. The incidence of PCD is estimated at 1/16,000 births.

The first cases, reported in the early 1900's, and characterized by a triad of symptoms that included chronic sinusitis, bronchiectasis and situs inversus, became known as Kartagener syndrome. Subsequently, patients with Kartagener syndrome, as well as other patients with chronic sinusitis and bronchiectasis, were noted to have "immotile" cilia and defects in the ultrastructural organization of cilia and so the term "immotile cilia syndrome" was used. The name was changed to "primary ciliary dyskinesia" later when studies showed that most cilia were motile, but exhibited a stiff, uncoordinated and/or ineffective beat PCD is genetically heterogenous and about 50 different geness have been described. Mutations in these genes account for 70-80% of cases of PCD. Inheritance pattern is mainly autosomal recessive but X-linked recessive inheritance is also reported. Our case has PCD – 19 caused by homozygous frameshift mutation in LRRC6 gene. It is expected that frameshift mutations cause more severe disease.

Treatment of PCD is mainly supportive and directed towards mucociliary clearance.

References

1. Gene reviews
2. OMIM

PATHOGENIC VARIANT CAUSATIVE OF THE REPORTED PHENOTYPE WAS DETECTED

Gene# (Transcript)	Location	Variant	Zygosity	Disease (OMIM)	Inheritance	Classification
LRRC6(-) (ENST00000620350.5)	Exon 5	c.630del (p.Trp210CysfsTer12)	Homozygous	Primary Ciliary dyskInesla, 19	Autosomal recessive	Pathogenic

Figure 2 : Results of WES

World's First Reported Case of Rhodoturula Infection Along with CMV Meningitis in a 2-Month-Old Treated at Fortis Noida

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Introduction

Postnatally acquired cytomegalovirus (CMV) commonly acquired via breast milk, develop symptoms of CMV infection in preterm or low birth weight infants. [(1)] Meningitis is a rare clinical manifestation of CMV infection and diagnosis of CMV meningitis is challenging. Molecular multiplexing platforms like biofire Filmarray panel detects multiple pathogens simultaneously and have excellent sensitivity and specificity. [(2)] Rhodotorula initially considered as an environmental saprophyte has now emerged as an opportunistic pathogen in the past few decades and can cause invasive infections in immunocompromised patients. Fungemia is the most reported infection with Rhodotorula followed by brain infections. [(3)] Coinfection of CMV and Rhodotorula has never been reported, here, we report a case of CMV meningitis and Rhodotorula Coinfection in a term infant which is first to our knowledge.

Case Report

A 2 month 10 days old boy presented to emergency department with high-grade fever (101 degree), increased irritability for 1 day and 2 episodes of seizures. The child was exclusively breastfed on the clinical findings, a provisional diagnosis of meningitis was kept, and further workup, including lumbar puncture and neuroimaging, was planned. Initial investigation has been summarized in [table 1.] Magnetic resonance imaging of brain showed diffuse extensive leptomeningeal enhancement along supra as well as infratentorial brain suggestive of leptomeningitis with prominent ventricles. He was managed conservatively with intravenous antiepileptics (levetiracetam), broad spectrum antibiotics and other supportive care. The patient had no repeat seizure episodes but there were persisting fever episodes. CRP again started rising at 56.1 and 88.6. Biofire Filmarray meningitis/encephalitis panel (BioMerieux, Utah) detected cytomegalovirus (CMV) in CSF. [(Table 1)] High levels of CMV (44638 copies/ml) in blood were also found in the blood. Considering meningitis with persistently high-grade fever spikes, loose stools, and CMV viremia, diagnosis of Disseminated CMV disease was, and ganciclovir was started. Simultaneously CSF culture also grew Rhodotorula mutants. Liposomal amphotericin B was also started, and broad-spectrum antibiotics were stopped. Breastfeeding was stopped and replaced with formula feed. Further investigations were sent to rule out any immunodeficiency. Human immunodeficiency viral load was not detected. The detailed immune work is normal. On systemic ganciclovir and liposomal amphotericin B, fever subsided, the child became clinically better and inflammatory markers CRP also started declining. [Table 2)] Antifungals were stopped after 4 weeks and antivirals ganciclovir were stopped after 6 weeks. On follow-up, after 3 months the child is doing fine with all milestones achieved in time.

Table 1. Initial Investigations on Admission

	19/01
Blood sugar	89 mg/dl
Haemoglobin	8.3 g/dl
Total leukocyte count 1000/cumm	3000
Platelet count	2.19 lakh/mm
C- reactive protein (CRP)	250.6 mg/l
Serum calcium mg/dl	8.9 mg/dl
Blood culture and Urine cultures	Sterile
Kidney function test and Liver function test	Within normal limits

Table 2. CSF analysis

	Initial episode	After 25 days	After 4 weeks of 2nd CSF
Total cell count cells/cumm	213	146	31
Neutrophil %	17	30	
Lymphocytes %	53	45	88
Mononuclear cells %	30	25	12
Protein mg/dl	386.5	101.4	35.7
Glucose mg/dl	8.9	31.1	34.4
Red cells cells/cumm	90	100	02
Bactec culture	Sterile	Sterile	Sterile
Fungal culture	Not done	Rhodoturola spp grown after 1 week	Sterile
Biofire	Not done	CMV detected	No pathogen detected

Discussion

CMV infection in newborn may be acquired in utero or via transplacental route known as congenital CMV infection. CMV can also be acquired in the immediate postnatal period (pCMV infection) via breast feeding probably occurs from a transmucosal route. CMV reactivation in breast milk occurs in ~90% of seropositive lactating women. [4] pCMV infection in full-term infants is of little consequence due to transmission of protective maternal antibodies which start about the 29th week of gestation. [6] However, symptomatic CMV disease can occur preterm infants or very low birth weight infants with varied clinical manifestations such as thrombocytopenia, sepsis, hepatitis, anaemia, lymphocytosis, cholestasis, pneumonitis, colitis, and meningitis [1]

Our patient presented with symptoms of neurological infections at 2 months of life, baby was clinically well after birth, so it is presumed that current CMV meningitis is due to post-natal acquired CMV probably via breast feeding as child was exclusively breast fed. However for a child who was born at term with adequate birth weight without any predisposing risk factor, diagnosing CMV meningitis was a real challenge. Initial CSF picture was suggestive of bacterial meningitis hence CMV was not considered and child was empirically treated with broad spectrum antibiotics but due to persistent clinical symptoms of fever, loose stools and transaminitis, alternative differentials for atypical and rare pathogens were thought of and detailed investigations were sent. Biofire film array is rapid multiplex testing platform that can detected wide arena of pathogens with a very short turn- around time. A recent meta-analysis have reported that Filmarray Biofire panel for meningitis/encephalitis has a sensitivity of 90% and a specificity of 97% for detecting microorganisms.[2]There are currently no clear guidelines regarding the criteria and duration of treatment for postnatally acquired CMV. We treated the child with ganciclovir. However, one week later CSF fungal culture grew Rhodotorula spp.

Rhodotorula spp is a pigmented yeast with low virulence. Earlier considered as non- pathogenic, it has emerged as an opportunistic pathogen in past few decades as it is notorious to cause invasive infections in immunocompromised patients [9]. Presence of central venous catheters, immunosuppression, transplant recipients, recent antimicrobial treatment, total parenteral nutrition, neutropenia, recent surgery are being implicated as important risk factors in patients with Rhodotorula fungemia. [9, 10] Nine cases of brain

infections in adult patients related to prolonged use of antibiotics and hospital stay have been reported in Indian study. [11]

In our case, Rhodotorula spp was identified in CSF by conventional fungal culture methods after one week, which was confirmed to be Rhodotorula mutans by MALDITOF. Our patient did not have any predisposing risk factors for such notorious infections; however child did have received broad spectrum antibiotics before diagnosis of Rhodotorula meningitis was managed in intensive care unit. We treated our patient with Amphotericin B for 4 weeks considering good penetration in brain for 4 weeks and after 4 weeks repeat CSF cultures were sterile.

Considering dual rare pathogens causing meningitis without any obvious predisposing risk factor, we speculated some kind of immunodeficiency, this led to further workup for immunodeficiency. The whole genome sequencing for primary immunodeficiency panel was done on blood samples. Two variants of undetermined significance were identified.

To the best of our knowledge, this is the first case reporting coinfection of CMV and Rhodotorula meningitis in a term neonate. This case highlights the important message for keeping suspicion of unusual pathogens in symptomatic infants not responding to the empirical broad-spectrum antibiotics as biochemical parameters could be misleading. This case also provides merits to the adoption of rapid multiplex diagnostics techniques in clinical practice.

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Coagulopathy and its Correction by Vitamin K in Children with Celiac Disease

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Background

25 % of children with Celiac disease (CD) have coagulopathy at the time of presentation. Diagnosis of CD involves endoscopy and multiple duodenal biopsies. Risk of bleeding with endoscopy and biopsy is more if there is underlying coagulopathy.

Objective

To study the correlation of coagulopathy with grade of histology in CD and to assess the response of a single dose of vitamin K on coagulopathy in children undergoing upper GI endoscopy and duodenal biopsy for CD.

Method: Study Design

Non-randomized interventional study.

Children (< 18 years) suspected to have CD referred for duodenal biopsies were prospectively recruited in study.

During the first 6 months (Group A) Prothrombin time (PT) was tested prior to endoscopy. During the next 6 months (Group B) children were given one dose of Vitamin K (5mg IM in <10years and 10mg IM in >10 years) 24 hour prior to endoscopy and PT was tested prior to endoscopy as in group A. A cut off INR of >1.4 was labelled as abnormal (coagulopathy). Subsequently PT/INR was compared in both the groups and correlated with severity of histology.

Results

Of 133 recruited children, 100 had confirmed CD by histology and were analysed subsequently. Both groups (A and B) had 50 subjects in each. The male female ratio in CD was 1.6:1 and the mean age was 5 years and 4 months. Group A and B were identical in terms of degree of demography and histological abnormality. Coagulopathy was seen in 32 % of children in group A and 14 % of children in group B and the difference was statistically significant. More than 50% of subjects with coagulopathy had advanced (Marsh grade IIIc) histology in both the groups which was significantly higher than those who had no coagulopathy. None had any significant bleeding during the endoscopic procedure in the study population.

Conclusion

CD with coagulopathy at presentation predicts advanced histological Marsh grade on duodenal biopsy. Coagulopathy can be significantly improved by single dose of parenteral vitamin K administration a day prior to endoscopy.

Keywords: Celiac disease, Coagulation, Children, vitamin K

Table I: Demographic characteristics of CD children in Group A and B

		Group A (n=50)	Group B (n=50)	Total (n=100) (%)
Place of patient recruitment	OPD	50	49	99 (98.5%)
	IPD	0	01	01 (1.5%)
	Total	50	50	100(100%)
Mean age at presentation (95% CI)		4.9±3.0 years (59.8±36.2 months) (49.51 - 70.10)	5.8±4.6 years (69.8±55. 9months) (53.94-85.73)	P value-0.29
Range(month)		**13-192**	**12-192**	
Sex	Male	31	29	60 (60%)
	Female	19	21	40 (40%)
	Total	50	50	100
Male to Female ratio		**1.6:1**	**1.4:1**	

(OPD- out patient department, IPD- in patient department)

Table II – Comparison of PT / INR in CD children between Groups A and B

INR	Group A (n=50)	Group B (n=50)	P value
INR<1.4	34	43	0.032
INR>1.4	16	7	
Total	50	50	

INR- International Normalised Ratio

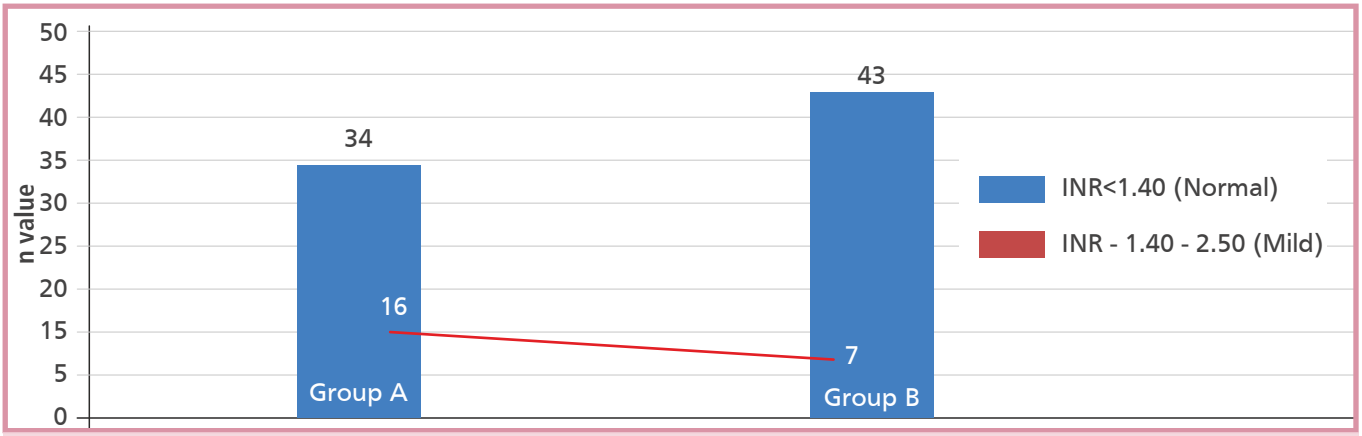

Figure 2 : Bar diagram illustrating the decrement in the numbers of children with deranged PT/INR after injecting vitamin K

Table III - Clinical features and lab parameter of CD in low vs high degree villous atrophy

Clinical feature	Low Degree villous atrophy (%)	High Degree villous atrophy (%)	Total (%)	p value
Chronic diarrhea	7 (16)	36(84)	43 (100)	0.27
Abdomen distension	9 (14)	55 (86)	64 (100)	0.11
Failure to thrive	12 (16.4)	61 (83.6)	73 (100)	0.13
Short stature	11 (17.7)	51 (82.3)	62 (100)	0.48
Abdominal pain	9 (25)	27 (75)	36 (100)	0.30
Pallor	14 (19)	60 (81)	74 (100)	0.79
Cheilitis	1 (33)	2 (67)	3 (100)	0.72
Frontal bossing	1 (16.6)	5 (83.4)	6 (100)	0.61
Hepatomegaly	0 (0)	11 (100)	11 (100)	0.18
Deranged PT/INR	12 (24)	28 (76)	50 (100)	0.012

(Low degree villous atrophy - Marsh grade 2 and 3a, High degree villous- Marsh grade 3b and 3c)

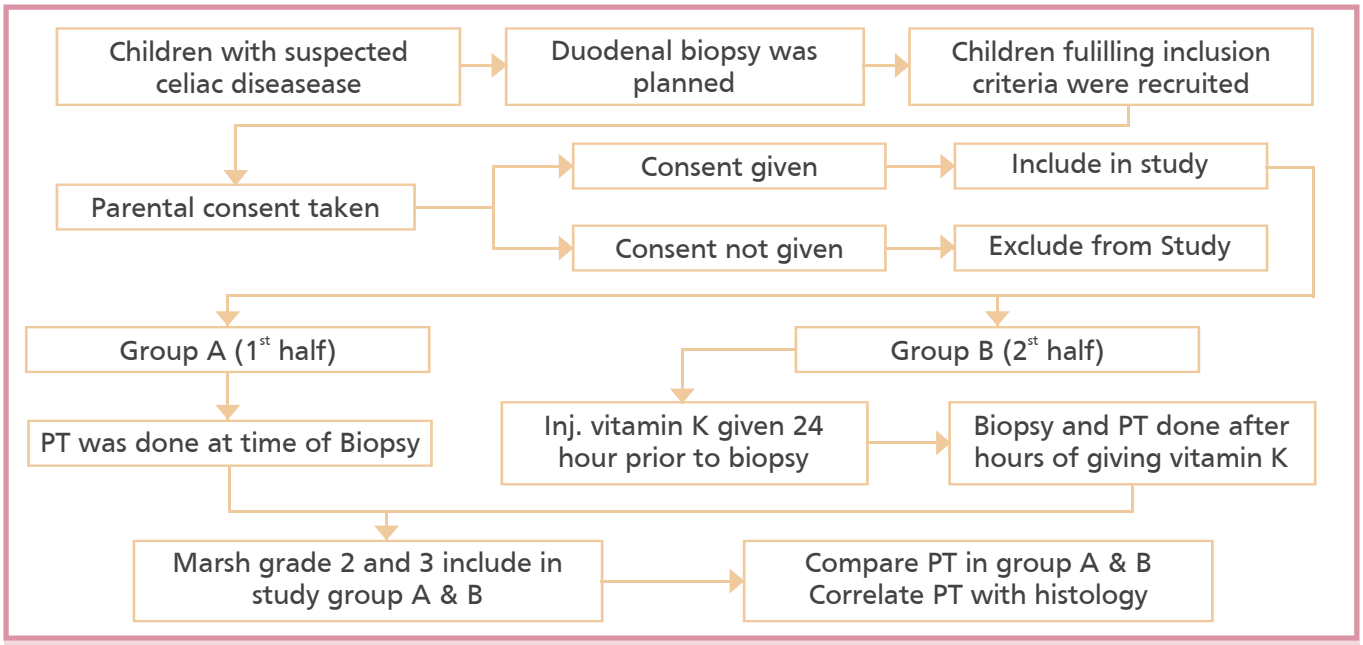

Figure 1 : Study Flow Diagram

Small Actions, Big Impact - Averting Iatrogenic Injury Among Neonates on Nasal Continuous Positive Airway Pressure (nasal CPAP) Therapy

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Introduction

Preterm neonates have a higher risk of developing respiratory insufficiency compared to term-born neonates. Respiratory support for neonates can be given through invasive ventilation and non–invasive ventilation. Non-invasive ventilation is preferred above invasive ventilation to prevent ventilation-induced lung injuries. Among all non-invasive ventilation devices, application of Nasal CPAP has been proven effective to achieve optimal gas exchange.

Nasal CPAP is the application of positive pressure to the airways of spontaneously breathing neonates throughout the respiratory cycle. Nasal CPAP is a relatively simple and effective therapy for respiratory distress syndrome when used in the neonatal intensive care unit (NICU). It involves use of an appropriately sized interface with a complementing head gear.

Nasal CPAP can be delivered either via nasal mask or binasal prongs. However, studies have described the development of pressure injuries on the nasal skin caused by these devices. The combination of an underdeveloped skin (in neonates) and the longer dependency on respiratory support can explain why nasal Pressure Injuries are frequently observed in neonates admitted in NICU. It manifests itself in following stages- Stage I: non-blanching erythema, on an otherwise intact skin; Stage II: superficial ulcer or erosion, with partial thickness skin loss; Stage III: necrosis, with full thickness skin loss.

Background

At Fortis Mulund NICU, routine nursing care aimed at preventing nasal Pressure Injuries (PI) mainly consisted of choosing the appropriate size nasal mask/binasal prongs/ head gear and frequent skin assessment. Despite these measures nasal Pressure Injuries- an iatrogenic complication, was a chronic concern.

Among 121 neonates who were admitted to the NICU from Jan 2020 to Sept 2021 there were 18 who required nasal CPAP therapy (14.88%). All the 18 neonates were born preterm and all of them developed either stage I or stage II nasal Pressure Injuries, with one instance of stage III Pressure Injury.

Objectives

- Retrospectively analyze the nasal Pressure Injury events and identify root causes for the same.
- Implement measures to reduce the nasal Pressure Injury Incidence rates.
- Evaluate effectiveness of the measures implemented in reducing the Pressure Injury Incidence rates.

Methodology

A team was put together with the NICU in-charge as the team-lead to deliberate on the recurring nasal pressure injury events and perform a literature review on measures to prevent it. Limited literature was available on preventive measures among neonates with use of nasal CPAP. As the root cause was identified as pressure, friction and sheer over the nasal septum, the team brainstormed and implemented initiatives in 2 phases.

Phase 1 (Jan 2021 onwards): Pressure relief via application of infant nasal CPAP seal (hydrocolloid material) was initiated to provide a protective barrier between the cannula and the neonates delicate skin while securing the cannula in place and providing a consistent seal, reducing the need to upsize prongs due to nasal leak. This strategy brought about reduction in the nasal Pressure Injury incident rates.

Yet, despite the use of the hydrocolloid seal, there was one incident (Sept 2021) of stage III nasal pressure injury reported that led to nasal septum damage in a neonate that was beyond repair. This triggered the need for a more robust mechanism to reduce the nasal pressure injury rates and hence the next phase of study was initiated.

Phase 2 (Oct 2021 onwards):

Brain storming was done by the team members to deliberate on additional measures to address pressure, friction and sheer on the nasal septum. The following strategies were then employed among neonates on Nasal CPAP: (i) Traction on the nasal tube was reduced by placing an elevated support to Venti circuit (ii) Infant nest was elevated at foot end to prevent the infant from sliding down the bed and thereby preventing the tube getting pulled, (iii) Two hourly pressure relief protocol (part of cluster care) initiated- includes release of cannula, skin assessment around the nasal orifice and application of moisturizer as needed.

Results

With the use of Nasal CPAP seal in Phase 1 there was reduction in the neonatal nasal Pressure Injury incident rates from 84.62% (Jan- Dec 2020) to 60% (Jan- Sept 2021).

Phase 2 interventions led to further drastic reduction in the nasal pressure injury rates (5.13%), with 2 instances of stage I redness which was captured and treated on time (Oct 2021-Mar 2023). Total 15 preterm babies on Nasal CPAP could be discharged without any nasal pressure injury till date.

Discussion

Simple strategies employed targeting on prevention of pressure, friction and sheer on the nasal septum with use of Nasal CPAP among neonates helped preserve the skin integrity on the nasal septal region as well as prevented long-term cosmetic sequelae which would have led to specialized follow up and possible surgery.

Conclusion

Preterm and low birth-weight neonates are prone to develop skin injuries due to the thin and unripe stratum corneum of the epidermis. The neonate's skin is one of the most underdeveloped organ systems at birth which, when admitted to the neonatal unit, becomes exposed to many therapeutic interventions and medical devices while it is not yet physiologically prepared. Along with routine skin care, customized special interventions will prevent serious complication among new-borns.

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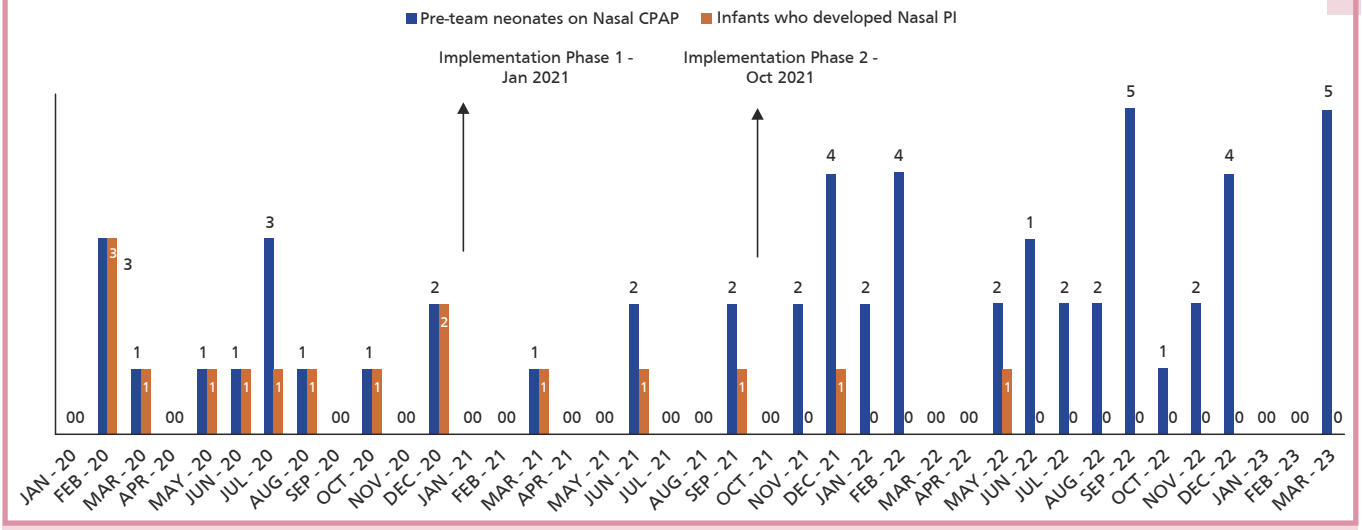

Nasal PI incident rate viz-a-viz pre-term neonates on Nasal CPAP

Neonatal Hypoglycemia: Fear or Fight?

(Neonatal hypoglycaemia- Screening and Management in roomed-in high-risk new-borns)

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Introduction

Neonatal hypoglycemia is a low blood glucose concentration in the first few days of life, it is a common metabolic disorder affecting newborns. It poses a significant challenge in the early neonatal period and can lead to both acute and long-term complications if not promptly recognized and treated. The delicate balance between glucose production, utilization, and regulation is particularly crucial in the vulnerable neonatal population, where energy requirements are high and glucose reserves are limited. The incidence of neonatal hypoglycemia varies widely, ranging from 1% to 15% of all newborns, depending on various risk factors such as maternal diabetes, prematurity, intrauterine growth restriction, and perinatal stressors.

Background

With the introduction of New Nursing sensitive Nursing quality Indicator - Detecting early signs of Hypoglycaemia, Fortis BG Road NICU nurses keenly monitored and reported neonate's Hypoglycaemic events as part of nursing practice audit. During the period of Nov 2021 to April 2022 there were 30 (12%) reported instances of < 50 mg/dl and 27 (10.5%) instances of < 45 mg/dl in high-risk new-born nursed in Labour Delivery Recovery Postpartum (LDRP) Unit.

In babies with hypoglycaemia event – 43% had maternal gestational diabetes mellitus and Type 2 diabetes mellitus.

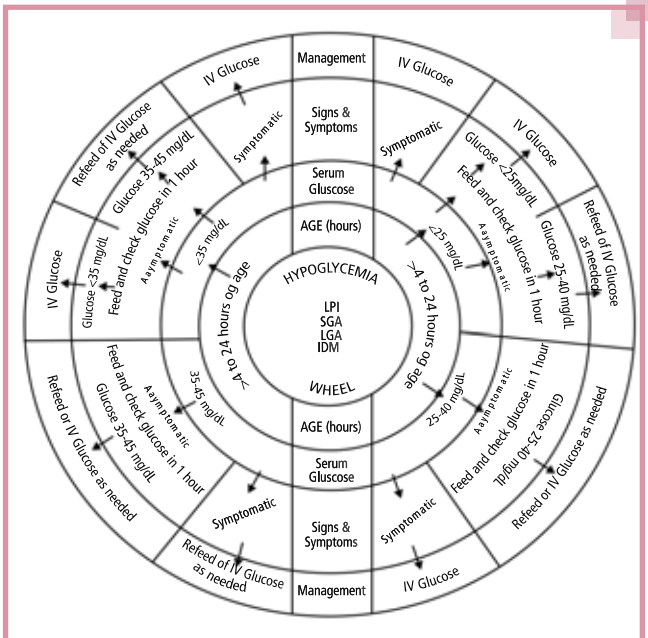
Objectives

- Implement measures to reduce neonatal hypoglycaemic events in high-risk new-borns nursed in LDRP
- Evaluate effectiveness of the measures implemented in reducing neonatal hypoglycaemia

Methodology

With the available retrospective data, a brain storming session on prevention of hypoglycemia in neonates was held with Neonatologists, obstetricians, lactation consultant and Nursing team. A revised algorithm of hypoglycemia management was adopted with reference to The American Academy of Paediatrics (AAP), Committee on Fetus and Newborn (COFN).

The criteria for hypoglycemia were changed to 45 mg/dl from the previous 50 mg/dl. While the data collection continued for both parameters.



Glucose Metabolism: Sugar Wheel Nomogram for Postnatal Glucose Homeostasis

Screening and Management of Postnatal Glucose Homeostasis in Late Preterm and Term SGA, IDM/LGA Infants

[(LPT) Infants 34-366/7 weeks and SGA(screen 0-24hrs); IDM AND LGA>34weeks (scrren0-12)]

Symptomatic and <40 mg/dL → VI glucose

ASYMPTOMATIC

Birth to 4 hour of age		4 to 24 hour of age	
INITIAL FEED WITHIN 1 hour Screen glucose 30 mintues after 1st feed		Continue feeds q 2-3 hours screen glucose prior to each feed	
Initial Screen<25 mg/dL		Screen<35 mg/dL	
Feed and check in 1 hour		Feed and check in 1 hour	
<25 mg/dL → IV glucose	25-40 mg/dL → Refeed/IV glucose as needed	<35 mg/dL → IV glucose	35-45 mg/dL → Refeed/IV glucose as needed

Target glucose>45 mg/dL prior to routine feeds

Glucose dose=200 mg/kg (dextrose 10% at 2mL/kg) and/or IV infusion at 5-8 mg/kg per min(80-100mL/kg per d). Achieve plasma glucose level of 40-50 mg/dL.

Symptoms of hypoglycemia include: Irritability, tremors, jitteriness, exaggerated Moro reflex, high-pitched cry, seizures, lethargy, floppiness, cyanosis, apnea, poor feeding.

Term neonates with the risk factors (Large and small for gestational age, maternal gestational diabetes, perinatal stress) and new-borns with symptoms of hypoglycaemia will be screened as per the algorithm.

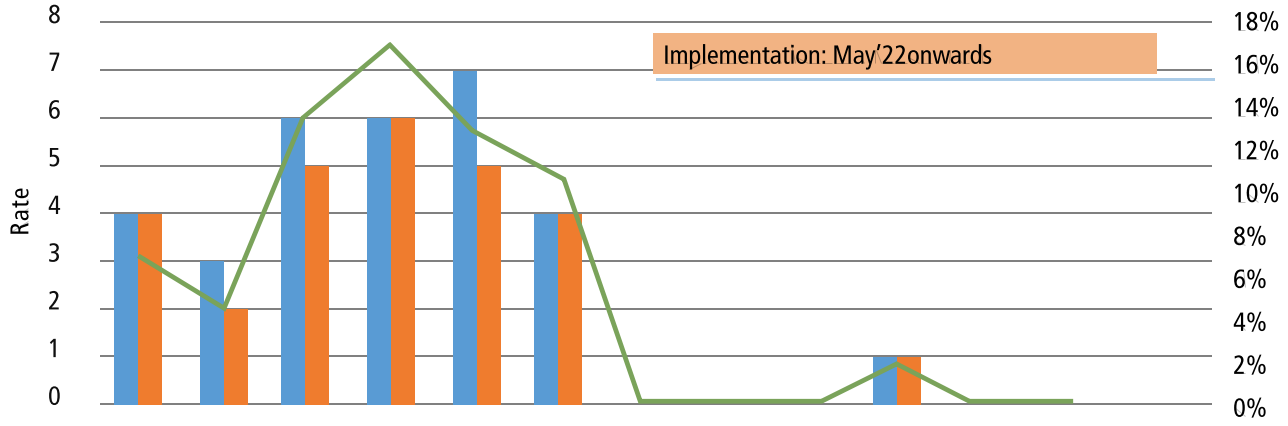
If GRBS reading < 45 mg/dl or shows a high reading a confirmatory lab test will be performed.

Results

The Number of Neonatal Hypoglycemia captured between Nov21 to April (Pre-Implementation) phase was ranging from 5% to 17% in the month of Dec21 and Feb22 respectively. The revised algorithm was implemented in May 2022, to screen and prevent hypoglycemia in high-risk newborns. Post-Implementation, the incidence of reported neonate hypoglycemia was 0% from May22 to Oct22 except for 1 reported incident reported in the month of Aug22.

Discussion

Total number of Neonatal hypoglycemia captured during the post- Implementation period was 1 in the month of August22 (Newborn with hypoglycemia within 2 hours of birth had no maternal GDM/DM. The baby had high TSH and required endocrinology consultation to evaluate the cause of hypoglycemia. The baby was discharged on day 5 of life). This reveals that after implementing and creating awareness to all the healthcare workers in NICU regarding the revised algorithm it was found that there was a marked reduction in the Neonatal hypoglycemic events.



	Nov-21	Dec-21	Jan-22	Feb-22	Mar-22	Apr-22	May-22	Jun-22	Jul-22	Aug-22	Sep-22	Oct-22
GRBS<50 mg/dl	4	3	6	6	7	4	0	0	0	1	0	0
GRBS<45 mg/dl	4	2	5	6	5	4	0	0	0	1	0	0
Rate	7%	5%	14%	17%	13%	11%	0%	0%	0%	2%	0%	0%

Conclusion

Neonatal Hypoglycemia is a preventable cause of brain injury. It is common, affecting 5-15% of all babies and approximately half of at-risk babies and is associated with a range of adverse sequalae. There has been significant reduction in the instances of newborn hypoglycemia due to early screening. The reduction is statistically significant Chi Square p value is <.05. While the protocol implementation has reduced instances of neonatal hypoglycemia, we shall continue to track causes and outcomes in the events identified.

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Rethink: Building Resilience and Support for Mental Well-being in Classrooms

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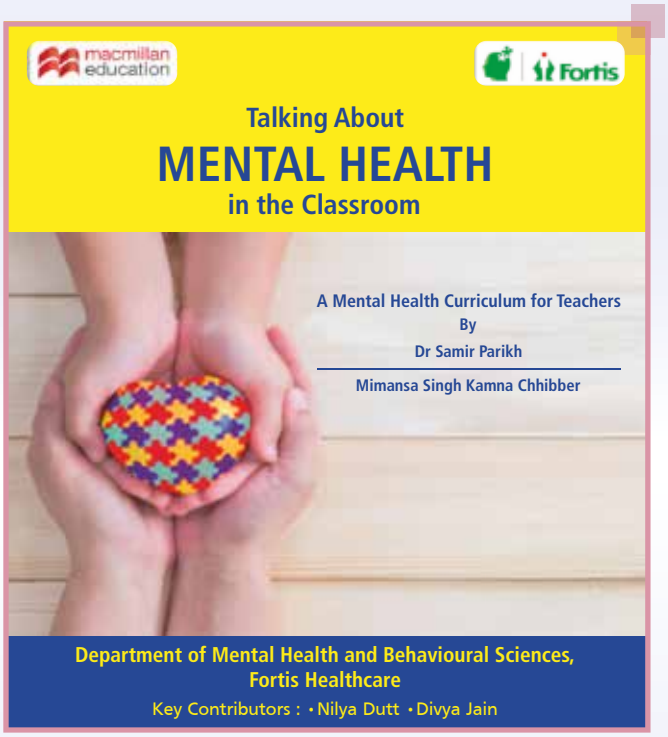
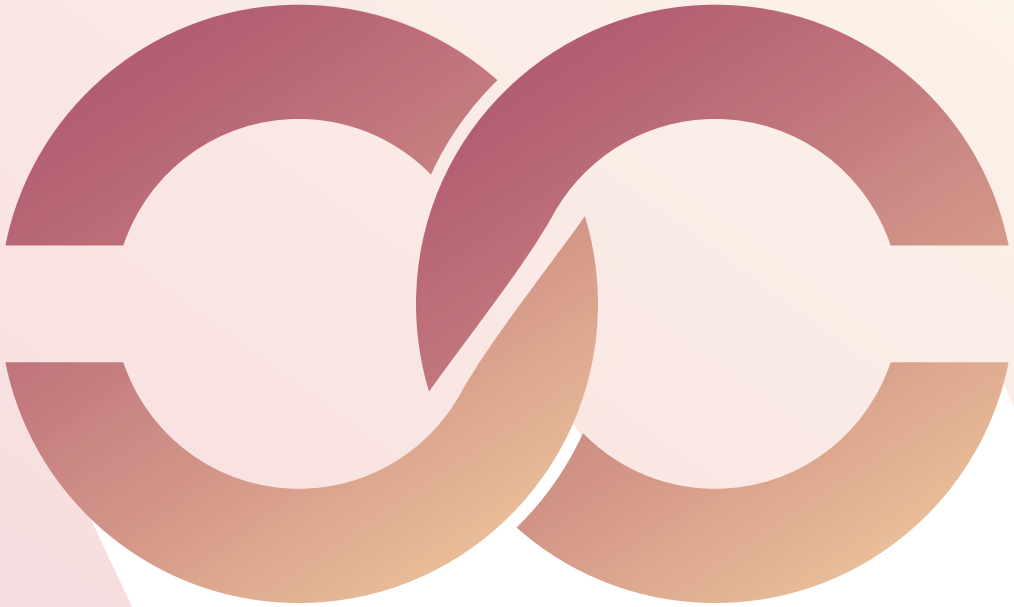
Did you know, that bullying is one of the leading risk factors for developing a mental health illness in later years? Bullying, peer pressure, substance use, academic pressures – these are just some of those things we take for granted as an unavoidable part of growing up.

Yet, when we think about mental health challenges, the image of children doesn't often cross our mind. What we don't recognize is that close to half of all mental health illnesses start by the age of 14 years. 1 in 7 young people between the age group of 10 - 19 years' experience a mental health disorder. Most of these go unrecognized and uncared for.

Stigma and the lack of awareness are the two greatest roadblocks to treatment when it comes to mental health. And so, it's time that we bring conversations about mental health into the classroom. Just like lessons on physical health and hygiene are a part of our textbooks in primary years, so mental health too must become a part of the curriculum.

The aim of the curriculum is not to go into the depths of every disorder, or turn every teacher into a counsellor – rather, it is to build understanding and empathy about mental health so children don't feel like they have to struggle alone and in silence. It's important to equip both students and teachers with an understanding of both mental health and mental illness to recognize red flags that can help in early identification and intervention. De-stigmatizing mental health by rethinking our social language and everyday vocabulary is also essential. With the right life skills, we need to make our children more resilient to bounce back from adversity and build a culture of support and well-being in the classroom. Most of all, we need to encourage children to ask for help when they're in distress, so they get the right care and support at the right time.





CLINICAL CONVERSATIONS

Recurrent Infections and Fever in Children - THINK BEYOND

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Introduction

Inborn error of immunity (Primary immune deficiency disorder) are rare disorders. The clinical presentation is variable and includes severe and unusual infections. Two cases presented here have an incidence of 2-3 million population (1st case congenital neutropenia) and 2nd case an incidence of 1 in 90000 male births (Agammaglobulinemia).

Case 1

A 4 year old female was admitted with fever for 2 months and multiple oral ulcerations for 7 days. But for mild pallor, clinical examination was unremarkable, with the child at 25th centile for weight for age. Initial investigations for prolonged fever revealed only low TLC (2910) with ANC (90). Broad spectrum antibiotics were started in accordance with protocol to treat febrile neutropenic patients. All investigations for viral/ bacterial / fungal infections were negative. Bone marrow examination was unremarkable, but the patient continues to show consistently low TLC and ANC. ELANE gene mutation was negative, G-CSF was started and the clinical condition of the child as well as TLC and ANC after G-CSF showed dramatic improvement.

Case 2

A 6 year old male child, was admitted with complaints of high-grade fever for five days, cough, purulent eye discharge, bilateral ear discharge and loose stools. On Examination, the child was 10th centile for age; bilateral crepitations on chest auscultation, haemorrhagic conjunctivitis, bilateral purulent ear discharge with cervical lymphadenopathy. Clinical picture was congruous with Adenoviral infection which was confirmed by respiratory bio-fire panel. Child was treated with broad spectrum antibiotics for secondary pneumonia (Chest X-ray showed right upper zone consolidation) and otitis media. The cause of continuous diarrhoea was investigated and the stool was found to be positive for Clostridium Difficale toxin which was treated accordingly. Blood, Urine and stool cultures were negative initially at the time of admission however a second Blood C/S grew Stenotrophomonas Maltophilia.

Going deeper into child history revealed that he had hospital admission and frequent infections requiring usage of multiple antibiotics, taking more than usual time for recovery. This lead to investigate for underlying immune deficiency. Immunoglobulin levels were very low for the child's age. Flowcytometry revealed absent CD19 cell line and normal T Cell line. Child was given IVIG and showed improvement.

Discussion

Congenital neutropenia is a cluster of genetic disease and causes low levels of neutrophils. These conditions can lead to recurrent infection of skin, sinuses, stomatitis and infection of lung, bone and nervous system.

Infections are difficult to eradicate and may persist for long. Neutropenia seen in these cases begins early in life and affected patients have ANC <1000, commonly <500. Genetic testing should be done for mutations and 50-60% have ELANE mutations, others may have HAX 1 and WASP mutations. Standard therapy includes infection of Granulocyte Colony Stimulating factor which can restore neutrophils.

Hematopoietic stem cell transplantation is potentially curative and should be considered in all patients.

Agammaglobulinemia is a rare inherited immunodeficiency disorder. This disorder is characterized by low or absent B cells, low or absent immunoglobulins. X-linked agammaglobulinemia is the commonest type. These children mainly present after 6-9 months, when maternal antibodies decline. Life threatening infections and chronic lung disease are the hallmarks.

Serum Immunoglobulin levels are low or absent. Flowcytometry will demonstrate low or absent B cells with normal T cells. Primary treatment is precaution against acquiring droplet infection, maintaining oral hygiene. IVIG is cornerstone of treatment. Hematopoietic stem cell transplantation is alternative modality.

Conclusion

Consider uncommon diagnosis in child with recurrent infections, infection by unusual organism and difficult eradication of organisms. Paying attention to small clues from lab reports can help arrive at early diagnosis.

Unseen is Eternal

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A 9-day-old preterm baby who was transferred from another hospital for laparotomy due to underlying diagnosis. The 10 days old male, born 32 weeks premature, with weight of 1.915 kg was brought to Fortis Hospital. Upon admission, weighed 2.3 kg and was diagnosed with NEC stage 2 and Pan-resistant Enterobacter/Klebsiella with multi-organ dysfunction.

History/Case

A preterm male baby was born prematurely at 33.3 weeks with LSCS, weighing 1.915 kilograms, and cried immediately after birth. There were no abnormalities found in the antenatal scans. The baby born in another hospital was transferred to the NICU on day 1 due to respiratory distress. The treatment initiated was continuous positive airway pressure (CPAP), intravenous (IV) antibiotics, and inotropic support. Initially, an umbilical venous catheter (UVC) was placed but later removed. Subsequently, a peripherally inserted central catheter (PICC) line was inserted in the left cubital fossa, resulting in thrombosis at multiple sites on peripheral veins.

Treatment Course in the Previous Hospital

In the previous hospital, the baby received iv paracetamol and enteral Ibugesic for hsPDA. Positive septic markers and meningitis were detected. Pan-resistant Enterobacter/Klebsiella were identified, treated with injection Colistin and Meropenem. On Day 9, the baby had abdominal distention; an intra-abdominal drain was inserted due to free gas observed in an X-ray. IV metronidazole and Flucon were given. Deterioration led to circulatory shock, prompting intubation with ET SIZE 3, FFP, and RDP administration. Fortis Hospital consulted for laparotomy and further management.

Outside Reports

HB	WBC	PLAT	BILI	CRP	OTHERS
16.9	10170	239			
14.9	11620	217	4.9		
15.3	10320	133	9.5 Ca-10, Na-137 K 4.3	24	
					CSF-No growth in culture CSF Routine-WBC-80, Lymphocytes-90%
12.7	3960	7000	7.8	216	Urea-88, Albumin-2.2, Na-128.9
12	7240	16000		207	Na-126, K-3.0

Treatment Course in Fortis NICU

Baby was maintained on Mechanical ventilation with 50% Fio2. PICC line was non-functional and the baby was in shock. Low UVC was inserted in emergency. Baby was maintained on a low dose of IV Adrenaline with IV Dopamine infusion with correction of hypoglycemia and electrolytes. IV Meropenem, IV Colistin were continued and in addition IV Penta globin was initiated due to the presence of two pan resistant bacteria and small for gestational age (SGA) with circulatory failure. IV albumin was administered as asymptomatic treatment to improve perfusion and address V/Q mismatch.

Test	Result
Hb	8.7
WBC	8.28
PLT	236
N/L	46/44
PT	14.1
INR	1.19
Na/K/Cl	133/4.51/102
CRP	9.8
Creatinine	0.31
Sr.bilirubin	11.3/9.91/1.39

Figure 1

On 2nd day of NICU admission, the baby underwent various tests. 2D ECHO revealed a structurally normal heart with no PDA or pulmonary Embolism (PE) and, normal biventricular function. USG cranium reported No intraventricular haemorrhage (IVH). USG abdomen showed a complex cystic lesion in right lobe of liver, gall bladder sludge, oedematous thickening of the stomach wall and small bowel loops and, mild ascites. **Paediatric surgeon reviewed the baby's clinical course and advised to continue conservative management and USG-guided tapping.**

At 20 hours of life: USG guided percutaneous tapping of liver abscess under IV Fresh Frozen Plasma FFP and iv Platelet coverage. The low umbilical venous catheter (UVC) was changed to IJV under IV FFP and iv platelet coverage. Over the next few hours, inotropes were weaned, and partial parenteral nutrition was initiated with gentle mechanical ventilation. After 48 hours of admission, the baby experienced a single episode of multifocal convulsion lasting approximately 5 minutes, which was terminated by an IV loading dose of levetiracetam. CT abdomen with contrast performed 24 hours later revealed a large abscess with air-fluid levels within the right lobe of the liver, accompanied by edema in the surrounding liver tissue. Both Blood culture and intrahepatic fluid grew Enterobacter cloacae, which was pan-resistant to all the antibiotics.

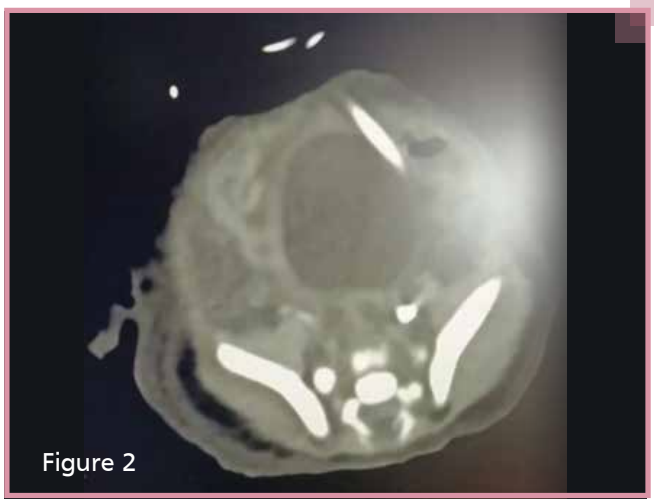

Figure 2

BLOOD CULTURE - BACTALERT AND SUSCEPTIBILITY

CULTURE; BLOOD - PRELIMINARY

CULTURE	POSITIVE
REMARK	GRAM STAIN : GRAM NEGATIVE BACILLI SEEN. BLOOD FROM LEFT DORSAL VEIN

CULTURE; BLOOD - FINAL

CULTURE	POSITIVE
ORGANISM	ENTEROBACTER CLOACAE COMPLEX

On the fifth day after NICU admission, a repeat USG-guided percutaneous drainage procedure was performed to drain the intrahepatic abscess. USG liver showed a large complex lesion in right lobe, with multiple internal reticulations, debris and echoes. Fluid from percutaneous drain showed presence ofyeast buds, hence started on Inj Fluconazole. The antibiotic regimen was modified as recommended by Dr. Kirti Sabnis, an infectious disease specialist. Aztreonam, ceftazidime + avibactam was prescribed while colistin was continued due to the presence of multidrug-resistant Enterobacter Cloacae.

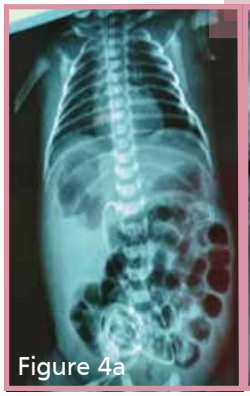
Figure 4a

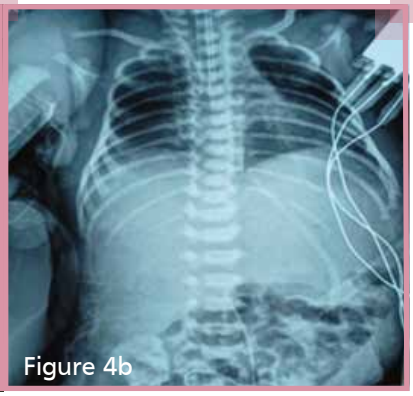
Figure 4b


Figure 5


Figure 6

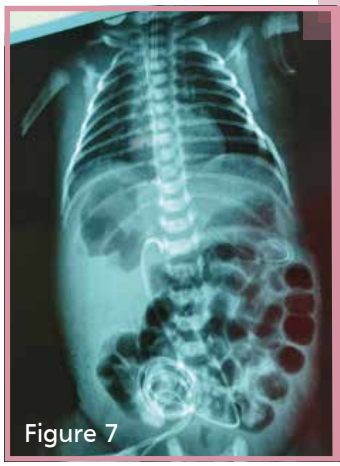
Figure 7


Figure 8

Nasogastric (NG) feeds were initiated after 5 days following the first aspiration, which baby tolerated well. Baby was serially monitored with X ray and ultrasound scans and laboratory tests, while providing necessary supportive care. On day 11 post- admission, baby was extubated, failed and hence re-intubated. However, after 48 hours, the baby was successfully extubated to non-invasive continuous positive airway pressure and transitioned to oxygen support followed by gradual weaning off. Further course was complicated by neonatal cholestasis with positive fungal markers. Thus, the baby received treatment with amphotericin B.

- Follow up USG abdomen on 35th day reported decreasing size of liver lesion (4cc), and partially distended urinary bladder with mobile internal echos (hence urine culture sent).
- Baby exhibited clinical activity, decreased icterus, and tolerated on-demand feeds. Switched from IV amphotericin B to enteral voriconazole due to decreasing levels of beta 3 glucan. and continued syrup septran. Baby was shifted to the ward, started breastfeeding, and discharged with oral medications including oral antifungals. Currently, at 9 months old, the baby is neurologically healthy.

Discussion

Neonatal Hepatic abscess in preterm infants is a rare entity with very high neonatal mortality and morbidity if left undiagnosed or untreated. But other way around, Neonatal pyogenic liver abscess, though rare, is associated with good outcome if diagnosed promptly and appropriate treatment instituted [1]

Neonatal hepatic abscesses are rare but should enter the differential diagnosis of a neonate with ongoing sepsis. This study serves to draw attention to their association with misplaced central (umbilical) catheters. Failure to respond to antibiotic therapy necessitates interventional drainage [2].

The combination of a carbapenem-containing regimen with colistin or MDR Enterobacteriaceae are often related to the production of extended-spectrum b-lactamases (ESBLs) and carbapenems-producing Enterobacteriaceae (CRE). [5]. Represent an increasing global threat. Carbapenem-sparing strategies should be used, when feasible, for ESBL infections. For treatment, combination therapy may be preferred over monotherapy for CRE[6].

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Ivermectin Poisoning - Report of Successful Management

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Ivermectin is a parasiticide and is the drug of choice in filariasis, scabies (crusted or if topical treatment has failed), and several other infestations [1]. It has a low rate of adverse reactions with toxicity occurring only with overdosing, resulting in adverse prognosis, as no specific antidotes are available [1,2]. Although rare in children, over 150 cases of serious neurological toxicities have been reported in adults [1]. We reported a case of ivermectin toxicity with encephalopathy, shock and aspiration pneumonia in a young child and its successful management [3].

A 6-year-old previously healthy girl (weighing 20.5 kg) presented with a history of accidental consumption of 60 mL of 1% w/v (600 mg) ivermectin lotion (30 mg/kg). She was undergoing treatment for scabies. After four hours of consumption, she had two episodes of vomiting followed by generalized tonic-clonic movements and loss of consciousness. On arrival, she was unresponsive, with Glasgow Coma Scale (GCS) 6/15, with tachycardia, tachypnea, hypotension and oxygen saturation of 79% at room air. Pupils were equal bilaterally (3 mm) with sluggish reaction. There were no signs of meningeal irritation. There was generalized hypotonia with absent deep tendon reflexes and fundoscopic examination was normal. Excessive salivation and bilateral crepitations were present. The child was admitted to the paediatric intensive care unit and intubated, ventilated, and given all the supportive measures including anticonvulsants for seizures and inotropes for hypotension. The National poison information centre was consulted and supportive management was advised as there was no specific antidote. Her urine output remained normal.

Patient started having high grade fever after 24 hours. Chest X-ray showed left upper lobe collapse and bilateral opacities along with leukocytosis and high CRP, for which appropriate antibiotics were given. In view of shock and hypotension, echocardiography was done, which showed normal left ventricular ejection fraction

of 65%.

Gradually, sensorium improved in the form of intermittent awakening after 48 hours, and GCS became 13/15 by day five. Patient became afebrile after four days. Hemodynamic improvement started only after day three, and vasopressors were slowly tapered off. Ventilatory requirements which were high initially, also decreased from day three and in view of neurological and hemodynamic stability, child was weaned off from ventilator on day five and extubated. Improvement. She was discharged after nine days of hospitalization in a stable condition.

Ivermectin, in standard therapeutic doses, has both excellent parasiticidal efficacy and high tolerability [1]. Our patient ingested 30 mg/kg of ivermectin, which was almost 100 times the recommended dose. In our patient, despite there being no specific antidote, vigorous monitoring, and supportive critical care treatment proved to be lifesaving.

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Conservative Management of Empyema - Complicated Broncho Pleural Fistula

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Post resection Broncho pleural fistula a constant and dreaded complication of pulmonary surgery, but rarely BP fistula can also complicate empyema and constitute a major therapeutic challenge.

Fistulas can develop spontaneously because of underlying pulmonary disease like Pulmonary tuberculosis, bacterial necrotizing Pneumonia or lung abscess.

We report a case of 3-year-old female with a history of severe dyspnea and fever 5 days before got referred. Child was treated with appropriate antibiotics for pneumonia, still the patient's condition worsened. After receiving at night CT chest done s/o left massive hydro-pneumothorax with Right mediastinal shift with left lower lobe collapse, left sided Intercostal chest tube was inserted but air leak persisted, repeat CT Chest was suggestive of persistent pneumothorax with Broncho pleural fistula. The persistence of pneumothorax in the patient depicts a possible Broncho pleural fistula which significantly increases his morbidity and mortality risk. Paediatric CTVS opinion was taken, conservative management and close follow up was advised. Over a period of week child responded well, ICDT removed, observed for 2 days in ward then discharged. Doing well on follow up.

Keywords : Necrotizing Pneumonia, Hydro pneumothorax, Broncho pleural fistula, complicated pneumonia, severe pneumonia, empyema

Intraventricular Haemorrhage in a Healthy Term New Born - A Case Report

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Introduction

Intraventricular haemorrhage (IVH) in new-born is usually a result of germinal matrix haemorrhage that ruptures through the ependymal lining into the lateral ventricles. Though it is commonly seen in preterm infants, it can occur in term infants. We report a case of IVH in an otherwise healthy term neonate.

Case Summary

Baby boy was born to 41 years old primi gravida with Type 2 Diabetes and Rh-negative pregnancy at 37+6 weeks gestation, by elective caesarean section (indication: breech presentation) with a birth weight of 2.7 kg. Baby cried immediately after birth with APGAR score of 8/10 at 1 min and 9/10 at 5 min. Baby had an uneventful postnatal period. On day 5 of life baby was admitted in NICU with lethargy, feeding difficulty and seizure (GRBS- 44 mg /dl). Baby was started on IV dextrose bolus, intravenous fluids and IV antibiotics. Baby had 2 more episodes of seizures in NICU and was started on injection Levetiracetam. Complete blood counts and CRP were normal. GRBS, serum electrolytes and serum calcium were within normal limits. CSF analysis showed elevated protein (110mg/dl) with normal sugar levels, normal CSF cell count (total count - 8, neutrophils 2, lymphocytes 6) and gram stain showed no bacteria. USG cranium showed intraventricular haemorrhage with mild prominence of right lateral ventricle. EEG showed diffuse asymmetrical slowing of delta activity in the left mid /posterior parietotemporal region and possible seizure activity in the left hemisphere. Coagulation profile showed slightly deranged PT – 20/INR -1.59. normal APTT –39.3, Fibrinogen levels -146. Protein C (23%), Protein S (51%) and Anti thrombin (65%) activity levels were reduced. Injection vitamin K was given. MRI brain showed intraventricular hematoma along dependent aspects of both the lateral ventricles and hypo intensities along right caudothalamic groove suggestive of haemorrhagic changes. In view of decreased protein c and protein s activity levels, Duplex scan of arterial and venous system of both lower limbs, USG abdomen with reference to portal vein and IVC and 2D ECHO was done which did not show any thrombus. Factor V Leiden mutation was not detected. Whole genome EXOME sequencing (PROC and PROS) reports awaited. New born metabolic screen was normal. TMS showed increase in glutamine, lysine and glutamic acid levels. Urine GCMS was normal. CSF, blood and urine culture showed no growth and antibiotics were given for 5 days. After 10 days of NICU stay baby was discharged in stable condition on direct breastfeeds with regular head circumference monitoring and cranial ultrasound on follow up.

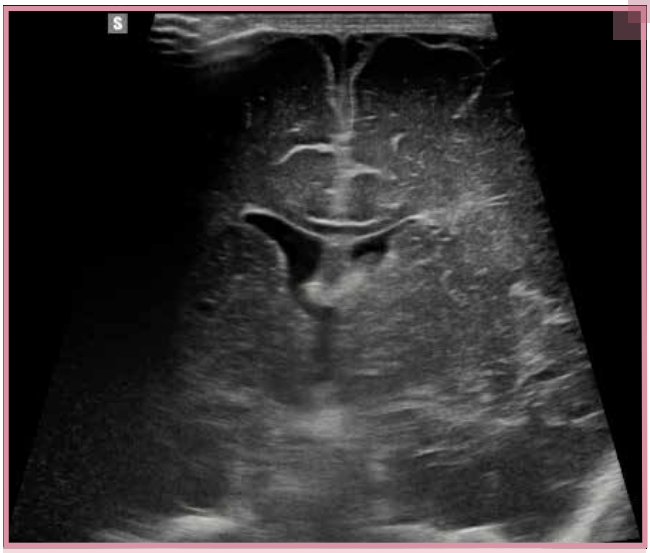

Figure 1 : IVH with mild prominence of right lateral ventricle

Discussion

The incidence of hemorrhagic stroke in term neonates has been reported as 0.17/1000 live births1. The majority of IVH is known to occur in the first 72 hours of life. Etiopathogenesis of IVH in preterm infants involves fragility of germinal matrix while in term neonates may include birth asphyxia, fetal distress, perinatal mechanical trauma, coagulation disorders and remains unclear in more than half the cases reported in literature. In a study by Afsharkas et al, 50% term neonates with IVH had coagulation disorders including DIC 6 (20%), sinovenous thrombosis (16.7%) and thrombocytopenia 4 (13.3%) of cases and protein C deficiency in one case3. Vitamin K deficiency is considered as a first possibility in any healthy term neonate with severe hemorrhage. Other causes include blood vessel malformations, cerebral sinovenous thrombosis and rarely protein S deficiency3. Sahriarian et al and Ayari et al have reported 2 cases of term neonates with severe PS deficiency4,5. Genetic factors including polymorphisms in the Factor V Leiden gene were associated with the atypical timing of IVH. Type and severity of symptoms (seizure, apnea, lethargy) is dependent on the size and location of hemorrhage, damage to the surrounding tissues and underlying predisposing factors of bleeding. According to Afsharkas et al, approximately 25 to 50% remained asymptomatic (IVH detected in imaging) and the source of bleeding was reported as choroid plexus hemorrhage (60%), germinal matrix (20%), parenchyma (6.7%). In our case, baby had no antenatal risk factors and had an uneventful natal and postnatal period and presented on day 5 of life. Work up for etiology ruled out dyselectrolemia, hypoglycemia, sepsis, metabolic causes. The possibility of coagulopathy secondary to protein c and s deficiency was considered and we plan to repeat activity levels after 3 months. PROC and PROS mutation analysis reports are awaited. Baby is being followed up at weekly intervals and is neurologically normal with evidence of mild ventricular dilation on cranial ultrasound study.

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TRIVIA

Crossword

Contributed by :
Dr Sukriti Sud
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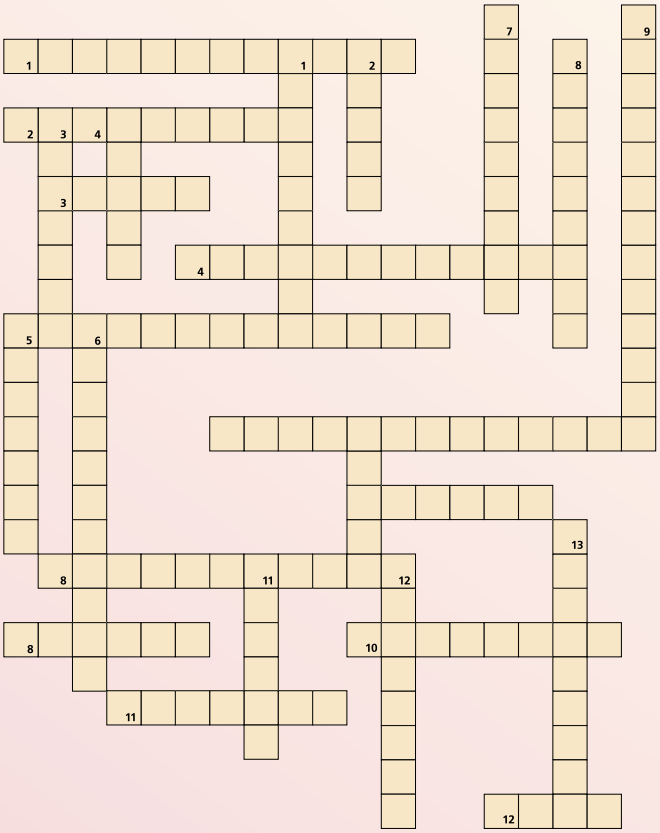

Across

1. A device to providing nutrient, fluid, and medication intravenously to children in a precise and controlled manner.(8,4)
2. Related to early childhood or infancy. (9)
3. Body parts with specific functions (5)
4. The process of administering vaccines to protect against infectious diseases. (12)
5. Pertaining to the growth and progress of a child's physical, mental, and emotional abilities. (13)
6. An infection of the small airways in the lungs, often caused by a viral infection and common in infants. (13)
7. Inflammation or infection of the ear, middle ear infection is commonly seen in children and often associated with ear pain and hearing loss. (6)
8. A medical instrument used by doctors to listen to a patient's heart, lungs, and other internal sounds.(11)
9. Describes a tendency to develop allergies or allergic conditions, such as asthma or eczema.(6)
10. synonym for the word 'adolescent' (8)
11. Viral infection causing a distinctive rash and high grade fever lasting upto 5 days in young children (sixth disease)(8)
12. A diagnostic technique in form of high-energy ionizing radiation that can pass through substances and create an image, commonly used in medical imaging to visualize the internal structures of the body (4)

Down

1. Born before the full term of pregnancy, often requiring specialized medical care (9 letters)
2. Viral infection characterized by swollen salivary glands, often affecting children. (5 letters)
3. A newborn baby, typically within the first 28 days of life.(7 letters)
4. A scoring system used to assess the health and well-being of a newborn shortly after birth. (5 letters)
5. Medical condition with specific characteristics (7 letters)
6. The act of administering a vaccine to stimulate the immune system against a particular disease.(11 letters)
7. Pertaining to a condition characterized by increased protein in the urine, low blood protein levels, and swelling (edema).(9 letters)
8. Respiratory infection causing inflammation of the lungs (9 letters)
9. A condition characterized by an accumulation of cerebrospinal fluid in the brain, resulting in an enlarged head and neurological problems.(13 letters)
10. A viral infection that causes swelling and narrowing of the upper airways, leading to a barking cough and difficulty breathing.(5 letters)
11. A high-pitched, wheezing sound that occurs during breathing and is often a sign of airway obstruction.(7 letters)
12. Involuntary bedwetting, typically seen in children who have not yet gained full control over their bladder during sleep.(8 letters)
13. A condition commonly known as "lazy eye," characterized by reduced vision in one eye due to abnormal development during childhood.(9 letters)

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Clinical Uses of Fructo-Oligosaccharides for Gastrointestinal Health in the Paediatric Population

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Abstract

Fructooligosaccharides (FOSs) are non-digestible carbohydrates that are one of the major classes of bifidogenic oligosaccharides. Studies find that prebiotics such as FOS display health benefits pertaining to but not restricted to the gastrointestinal (GI) tract of an infant. This review article aims to discuss the therapeutic potential of FOS for different paediatric gut conditions. FOS in varying concentrations has been found to prevent constipation and soften stools; reduce the incidence and severity of diarrhea; and alleviate GI discomfort symptoms such as vomiting and regurgitation. Infants and children seem to tolerate both short- and long-chain FOS molecules well. Although FOS is beneficial for infant and child health, there is still a need for rigorous clinical trials and long-term follow-up studies to understand if FOS supplemented in infancy can confer long-term effects in adulthood.

Keywords: Bifidobacterium, Fructooligosaccharides, Gut health, Infant, Inulin, Oligofructose, Prebiotics

In infant's gut is colonized at birth by different microbiota which is influenced by numerous environmental and host factors. Infant feeding type is a major factor influencing gut microbiota composition and good gastrointestinal (GI) function [1]. Apart from water, omega-3 lipids, protein, and micronutrients; breast milk is rich in oligosaccharides which exhibit prebiotic properties [2,3]. The International Scientific Association for Probiotics and Prebiotics defines prebiotics as a substrate that is selectively utilized by host microorganisms conferring a health benefit[4].

This unique human milk prebiotic component is practically absent in cow's milk [2,3]. Infant formulas routinely use plant-based prebiotics such as Fructooligosaccharides (FOS), galactooligosaccharides (GOS), or polydextrose to mimic the ones present in human milk[3].

This review will enlist different prebiotics, with a focus on FOS for the infant gut microbiome. It will also discuss the therapeutic potential of FOS for different paediatric conditions.

Conclusion

An infant's gut is colonized at birth with various microbiota by many factors playing a role. The abundance of certain bacterial taxa denotes good gut and overall infant health. FOS is one of the preferred prebiotics as it has been shown to promote the growth of two of the most beneficial gut bacteria, namely, Bifidobacteria and Lactobacillus in the infant.

The growth of these probiotic bacteria and their metabolites, namely, SCFAs have been identified as the main drivers of conferring protection against various gut-related conditions. Previously, reviewed studies have consistently found FOS to reduce the incidence and severity of diarrhoea; alleviate GI discomfort symptoms such as vomiting and regurgitation; prevent constipation and soften stools. When it comes to tolerance, both short- and long-chain FOS molecules seemed to be well-tolerated by infants and children without displaying adverse GI problems, as also noted by the ESPGHAN committee.

However, there are some limitations – most of the studies had different age groups and different dosages of FOS used. Most of the studies included in the review only identified the changes in the fecal microbiota during the period of supplementation. Long- term follow-up of the participants was not carried out.

The reviewed literature showed that FOS helps foster good gut bacteria development thus alleviating common gut-related conditions in the infant. However, there is a need for rigorous clinical trials and long-term follow-up studies to understand if FOS supplemented in infancy can confer long-term effects in adulthood.

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Successful Multidisciplinary Approach in Managing Complications in a Four-Year-Old Child with Extrahepatic Portal Venous Obstruction (EHPVO) - A Case Report

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Introduction

Extrahepatic portal venous obstruction although rare in children is a significant cause of portal hypertension in children which leads to significant gastrointestinal bleeding in paediatric age group. Portal hypertension may also lead to complications such as hypersplenism, ascites, hepatopulmonary syndrome and Porto-pulmonary hypertension that may require organ transplantation.

Case Summary

4-year-old girl child a known case of Extrahepatic Portal Venous. Obstruction (EHPVO) was brought from Mysore District. Child had undergone Upper GI Endoscopy which was suggestive of perforation at Gastroesophageal junction and was referred to Fortis Hospital for further management. Prior to this child was treated with endoscopic ligation and sclerotherapy for upper GI varices.

At admission, child was sick looking with Right ICD in situ with respiratory distress. She was started on oxygen via high flow nasal canula, empirical antibiotics and continued Nil orally. CECT chest showed mediastinitis right sided pleural effusion with? perforation at lower end of esophagus. She was taken up for thoracoscopic decortication and closure of the esophageal perforation. Post operatively she had massive episodes of requiring resuscitation with blood products and octreotide infusion. Her blood culture grew Burkholderia capecia, antibiotics with upgraded accordingly. She continued to have persistent anaemia, thrombocytopenia with fever spikes unresponsive to broad spectrum antibiotics. At end of second week it was decided to take her for splenectomy and Splenorenal shunt after discussion with ID specialist and Paediatric surgeon since she continued to have intermittent hematemesis and any endoscopic procedure was contraindicated due to perforated esophagus. child underwent Splenectomy and distal Spleno-renal Shunt surgery successfully. She was started on Heparin infusion post operatively. On POD 2 she developed massive hematemesis requiring extensive resuscitation and reversal of heparin with Protamine sulphate.

With suspected leak from mediastinal drain, she underwent repeat Thoracotomy with closure of esophageal rent and feeding jejunostomy. Her fever

spikes persisted, USG abdomen showed septated ascites for which she underwent exploratory laparotomy and closure of feeding jejunostomy by the end of 4th week. In view of persistent fever spikes at end of 5th week, PET scan done which was not suggestive of any focal lesion or collection. In view of last two consecutive cultures being sterile after 4 weeks of susceptible antimicrobial therapy, no focal collection on PET - CT and raised inflammatory parameters, diagnosis of Post infectious inflammatory process was thought off and child was started on oral steroid and IVIG after discussing with ID specialist.

Her fever subsided gradually over next 7 days. She remained afebrile for 4 days prior to discharge, was accepting oral feeds and hemodynamically stable. She was discharged on oral steroid with oral antibiotics. At subsequent follow ups her inflammatory parameters gradually normalized and was continued on oral steroids under cover of Antibiotics along with oral anticoagulants.

Discussion

EHPVO is the most common cause of prehepatic Portal hypertension in children.1,2 It could be consequence of neonatal omphalitis, catheterization of the umbilical vein or other intraabdominal infections or sepsis. EHPVO has also been reported as a consequence of perinatal events such as asphyxia, persistent pulmonary hypertension, sepsis and presence of congenital heart disease. Factor V Leiden and acquired JAK2 (JAK2V617F) mutation have been detected in adult patients with EHPVO 3,4. Nevertheless, none of these prothrombotic hereditary mutations have been found in children with EHPVO. Most of the paediatric cases of PHT associated to EHPVO are diagnosed at the age of 10-14 years. In contrast, our patient had confirmed Protein C and Protein S deficiency with very early presentation.

Life-threatening hematemesis and melena are the most frequent presentation with variceal bleeding as the most common cause. Complications such as hypersplenism, cholangiopathy, ascites, hepatopulmonary syndrome and Porto-pulmonary hypertension are associated with EHPVO in children. An unexplained pancytopenia associated with hypersplenism is an important clinical feature that should be recognized early.

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Childhood Malignant Rhabdoid Tumor - Case Series

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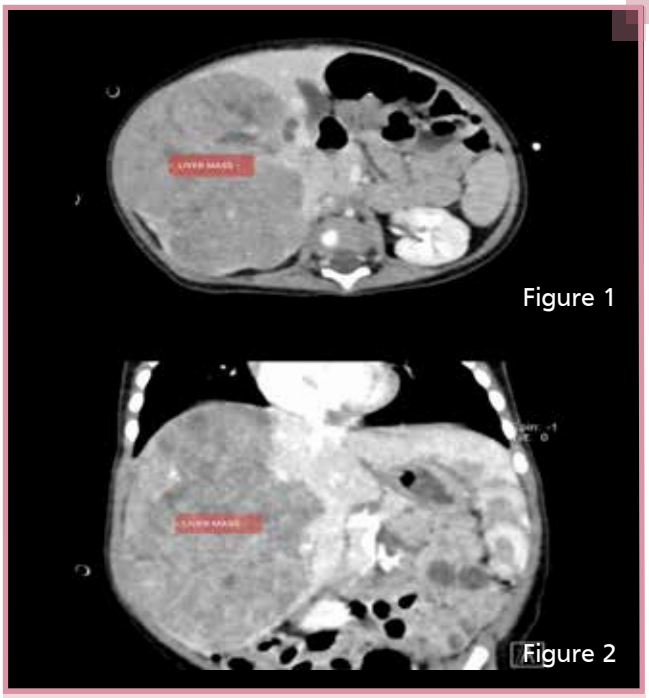
Introduction

Malignant rhabdoid tumors (MRT) are rare, aggressive malignancies occurring in infants and children less than 3 years of age. MRT usually arise in the CNS (~65%) (ATRT - atypical teratoid/rhabdoid tumor), but also in extracranial sites such as (~35%) in the kidneys (RTK – RT of the kidney) and other soft tissues (eg, liver).

Case 1

Seven-month-old baby presented with abdominal mass felt over the right side in the last 2 weeks. On examination, there was hepatomegaly. USG of the abdomen revealed hepatomegaly with heterogenous solid lesion in the liver. CT abdomen revealed a heterogenous mass measuring 8.7 x 7.2 x 6.6 cm, in the right lobe of the liver (Fig1and2).



Contrast enhanced CT scan of abdomen showing mass in the segments V, VI, VII, VIII of liver

The clinico-radiological diagnosis was hepatoblastoma. Serum Alpha-Fetoprotein was normal even after serial dilutions done to mitigate Hook effect. Hence, diagnostic liver biopsy was done. Microscopy demonstrated rhabdoid morphology; (Fig 3). On immunohistochemistry (IHC), there was loss of expression of SMARCB1 (INI-1) in the tumor cells suggestive of a malignant rhabdoid tumor of the liver; (Fig 4). Staging workup was negative for metastasis. Child underwent chemotherapy and surgery.

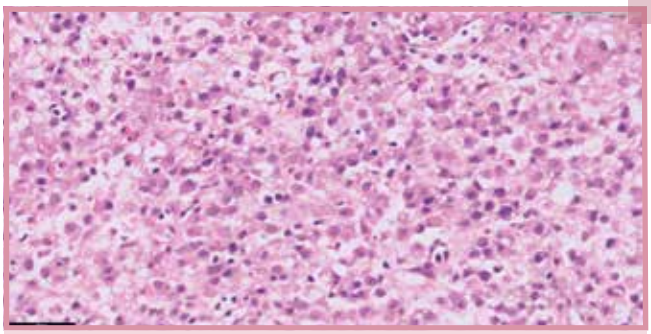


Figure 3 : Microscopy demonstrated sheets of small round tumor cells with abundant cytoplasm, eccentrically placed nuclei - rhabdoid cells

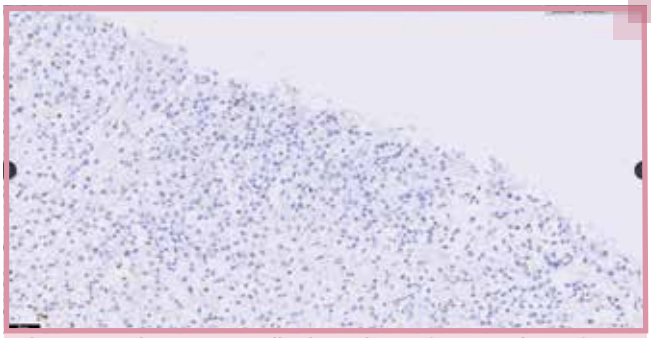


Figure 4 : The tumor cells show loss of expression of INI-1

Case 2

Nine-month-old baby presented with history of failure to thrive, fever and vomiting since 2 months. USG abdomen showed a mass in the left kidney. CECT abdomen revealed a 3.7 x 2.9 cm enhancing mass in the interpole region of the left kidney; (Fig5).

The clinico-radiological diagnosis was Wilms tumor. As the child had persistent vomiting, lethargy and a wide-open anterior fontanelle, MRI Brain was done. MRI showed a 6.1 x 4.2 x 3.6 cm posterior fossa mass occupying the fourth ventricle, compressing the aqueduct causing severe obstructive hydrocephalus with periventricular ooze; (Fig6,7and8).

Emergency resection of tumor was done and pathology showed malignant primitive embryonal tumor. On IHC, there was a loss of INI-1 protein expression suggestive of ATRT, CNS WHO grade 4. In view of synchronous tumors, blood sample was sent for germline testing to detect rhabdoid predisposition syndrome. Child was started on chemotherapy and planned for nephrectomy and adjuvant radiotherapy.

Figure 5

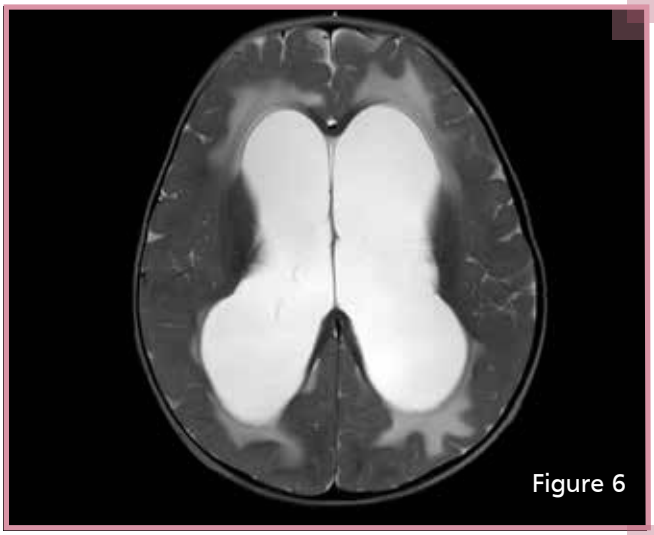


Figure 6

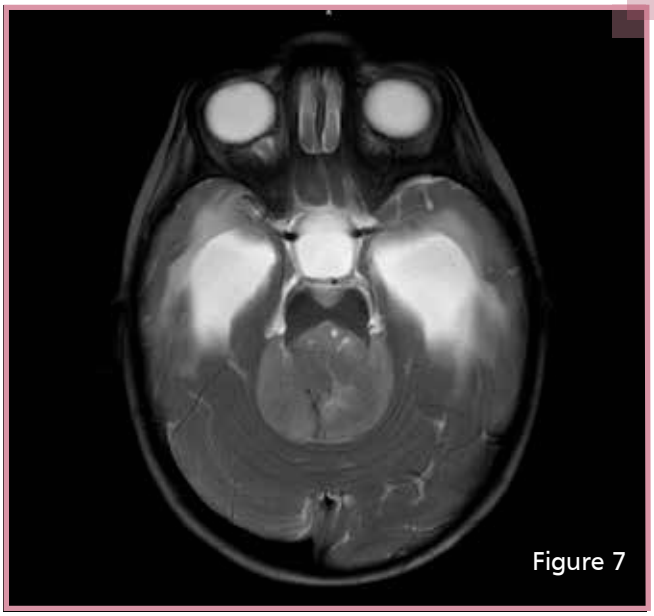


Figure 7

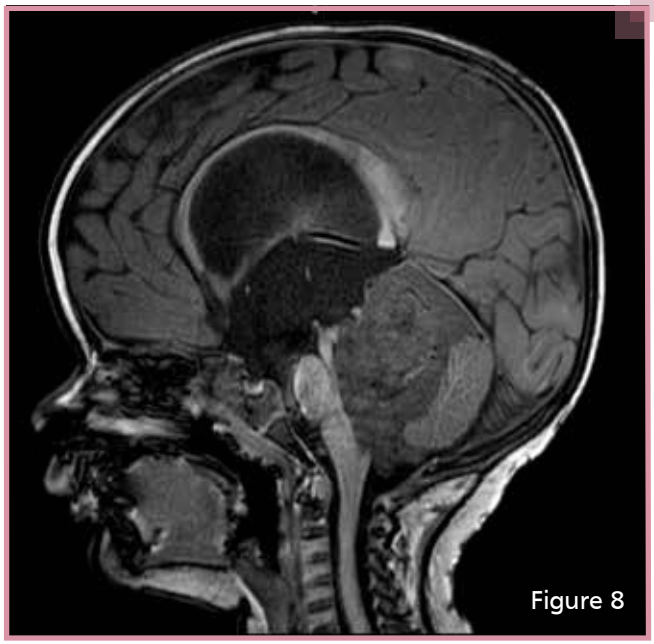


Figure 8

Discussion

Fewer than 40 cases of liver and synchronous MRT have been reported, hence the challenges in diagnosing these rare tumors. MRT should be kept as a differential diagnosis in early onset (<1 year) and metastatic brain, renal and liver tumors. As many as 10% to 15% patients with RTK present with synchronous ATRT and exhibit germline mutations in SMARCB1. Genetic testing and counselling are imperative in these cases for future pregnancy.

Acknowledgement

We thank Dr. Kunal Sharma, Associate Director and Section Head (Centre of Excellence and Histopathology), SRL Mumbai for his expert opinion on pathology and sharing quality images.

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Kikuchi Lymphadenitis: A Differential Diagnosis of Tuberculous Cervical Lymphadenitis

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Abstract

Patients from countries, endemic with tuberculosis, who present with febrile lymphadenopathy refractory to first line antibiotics are often empirically treated for extra-pulmonary tuberculosis. However, Kikuchi-Fujimoto Disease (KFD) or histiocytic necrotizing lymphadenitis, a self-limiting and benign condition, presents with similar clinical symptoms.

We present an adolescent with febrile lymphadenopathy, who was initially treated for tubercular lymphadenopathy, before a diagnosis of KFD was made.

Keywords : Kikuchi Fujimoto Disease, Febrile Lymphadenopathy

Case Report

A 16-year old Indian male initially presented with upper posterior cervical lymphadenopathy which resolved with a course of azithromycin. However, he returned after six months complaining of tender left cervical lymphadenopathy, fever, night sweats and weight loss, which persisted despite two courses of antibiotics. His oropharynx and tonsils appeared normal. He was prescribed anti-tubercular therapy based on his clinical history. However, his condition deteriorated after three weeks on this medication and he was taken to the emergency department. Laboratory workup revealed leucopoenia (3.83× 109/L) with an elevated erythrocyte sedimentation rate of (29 mm/h), C-reactive protein (68.6 mg/L), alanine aminotransferase (61 U/L) and aspartate aminotransferase (95 U/L). Investigations for tuberculosis (Mantoux skin test and CBNAAT performed on sputum sample), typhoid, CMV, EBV, malaria, dengue, leptospirosis and scrub typhus were all negative. His blood and urine cultures were negative. SARS-CoV-2 testing by rapid antigen detection was likewise negative, with no significant levels of SARS-CoV-2 IgG antibodies. Chest radiography and abdominal ultrasound scans yielded no significant findings. Two enlarged left cervical lymph nodes each measuring 2 × 2 cm were excised. Histopathological analysis showed, under low power, karyorrhectic debris in a background of paracortical foci of coagulative necrosis, and under higher power (Figure 1), crescentic histiocytes and histiocytes with foamy cytoplasm were observed within the necrotic areas. Acid fast bacilli, granulomas and neutrophils, were not detected. Immunohistochemistry was positive for CD68, CD20, CD3; and negative for PAX-5, CD15, CD30.

The patient was diagnosed with KFD and treated conservatively. His condition improved within 48 h, with complete resolution of symptoms during the following four months.

Discussion

Patients with KFD generally present with fever and tender unilateral cervical lymphadenopathy along with other non-specific symptoms including chills, arthralgia, night sweats, skin rashes, and weight loss. KFD can also manifest as mediastinal lymphadenitis, which might be misdiagnosed as tuberculosis.[1] The onset of KFD is acute or subacute, evolving over 2–3 weeks.[2] However, these symptoms can be present in many other conditions including tuberculosis, lymphoma, HIV, SLE and infectious mononucleosis.[3] Up to 40% of cases of KFD are mistaken for other causes of lymphadenitis.[4]

Availability bias amongst physicians who have repeatedly encountered a certain disease can lead to misdiagnoses of diseases with similar features.[5] In all cases of chronic lymphadenopathy, excision biopsy of the lymph node and its histopathological analysis is mandatory to rule out bacteriological causes and for the definitive diagnosis of KFD.[2]

Characteristic histopathologic findings in KFD include paracortical foci of coagulative necrosis with karyorrhexis and disruption of nodal architecture, crescentic histiocytes and macrophagic histiocyte with foamy cytoplasm. The major cell type is CD8+lymphocytes. Neutrophils are characteristically absent.[2] Granulomas and acid-fast bacilli are also absent, thereby ruling out extrapulmonary tuberculosis.

The absence of haematoxylin bodies and neutrophils rule out SLE.[4] In tuberculous endemic countries, patients are often empirically treated for extra-pulmonary tuberculosis based on clinical symptoms only. Prompt

histopathological examination can prevent misdiagnosis and harmful reactions due to unwarranted toxic therapy.

KFD is generally self-limiting and spontaneously resolves in one to four months. A recurrence rate of 3–4% has been reported.[2] There is no specific treatment but supportive therapy for symptomatic relief include anti-inflammatory drugs, prednisolone, chloroquine and hydroxychloroquine.[2]

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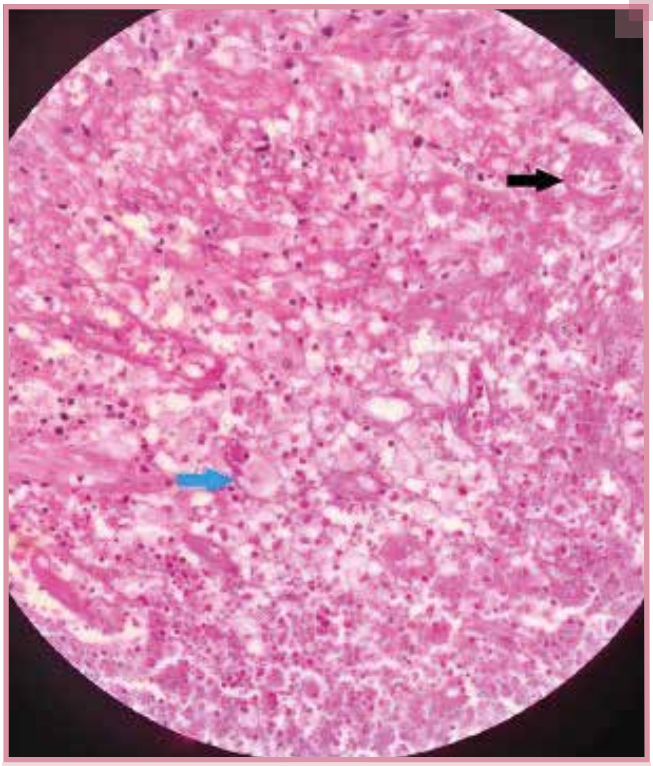

Figure 1 : Blue arrow pointing at crescentic histiocytes and black arrow pointing at histiocytes with foamy cytoplasm in areas with necrotic debris (HandE, 400×).

A Rare Case of Coronary Cameral Fistula in an Asymptomatic Child Where Clinical Examination Clinch the Diagnosis

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Introduction

Coronary fistula is defined as a connection between a coronary artery and a cardiac chamber or any segment of the systemic or pulmonary circulation. Coronary artery fistulas are the most common congenital anomalies of the coronary artery, representing nearly half of all coronary artery anomalies. It can be isolated or associated with other CHDs, such as tetralogy of Fallot (TOF), atrial septal defect (ASD), PDA, and VSD.[1]

Case Description

"A 2-year-old asymptomatic child presented in the OPD with a continuous murmur and suspicion of patent ductus arteriosus (PDA), but no PDA was detected on echocardiography. On examination, S1 was normal, S2 split 3/6, and there was a continuous murmur heard at the right sternal border. The echocardiogram revealed a dilated left main coronary artery (LMCA) and left anterior descending artery (LAD) with a Z score of +3.0. The interatrial septum (IAS) and interventricular septum (IVS) were intact, and there was no mitral regurgitation (MR), aortic regurgitation (AR), or PDA. The left atrium (LA) and left ventricle (LV) were mildly dilated, and turbulent flow was observed in the right ventricular (RV) cavity. As a result, a diagnosis of coronary cameral fistula to the RV was made.

Since the child was asymptomatic, the family did not agree to any intervention and instead started the child on aspirin. However, a follow-up echocardiogram revealed progressive dilatation of the LMCA, LAD, and an increase in LV dimensions. After 6 months of the initial evaluation, the family decided to proceed with closure. The patient was taken to the cath lab, where an aortic root angiogram showed a dilated LMCA and LAD, while the right coronary artery (RCA) appeared normal (refer to Fig 1). A selective left coronary artery (LCA) angiogram revealed a dilated LAD with filling of the RV seen through its distal communication."

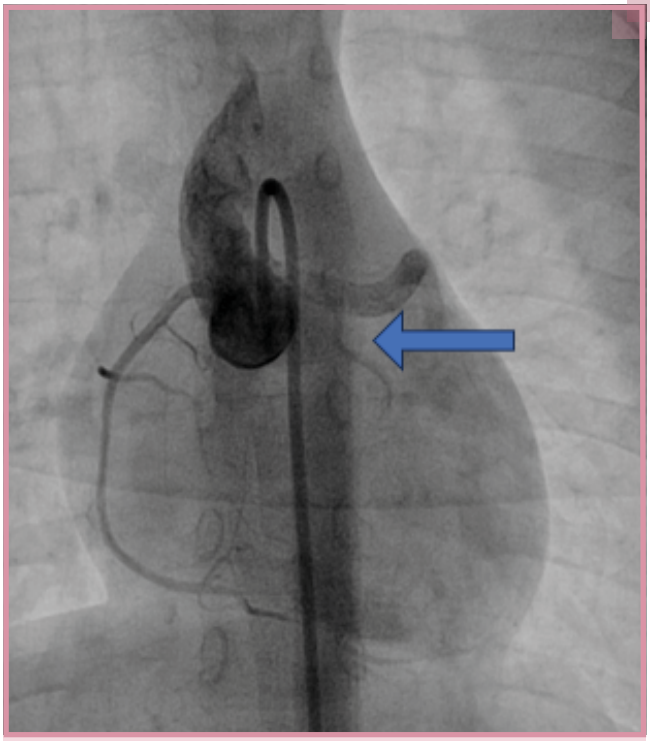

Figure 1 : Dilated LMCA , RCA normal

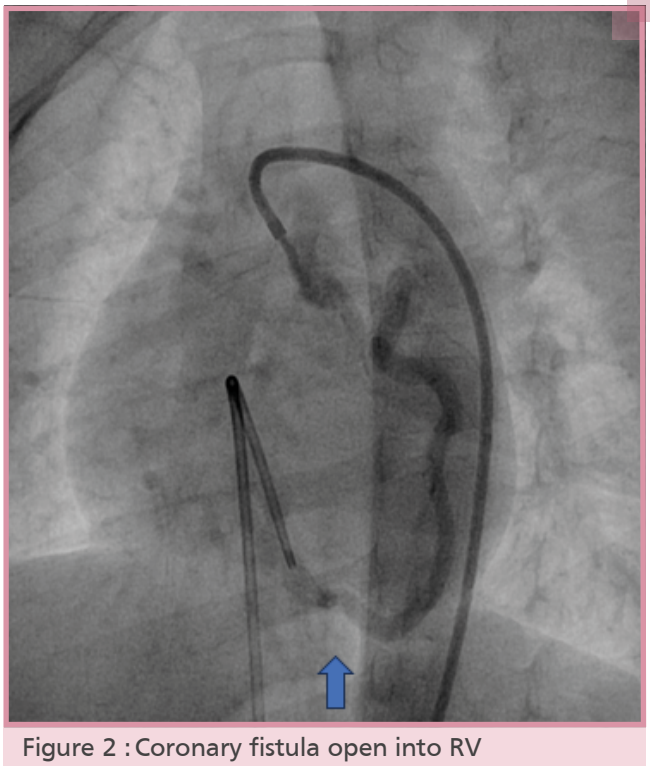

Figure 2 : Coronary fistula open into RV

Dilated left anterior descending coronary artery (LAD) measures 4.0mm, with its distal part communicating into the right ventricle (RV) through a narrow and tortuous opening measuring around 1.5mm. Catheterization data revealed a significant increase in blood oxygen saturation at the RV, with 89.2% saturation compared to 70.6% saturation at the superior vena cava (SVC). The fistula was closed with a 6/6 mm Amplatzer Vascular Plug under fluoroscopic guidance. The final angiogram showed that the vascular plug successfully occluded the distal LAD with no filling of the RV(Figure 3).

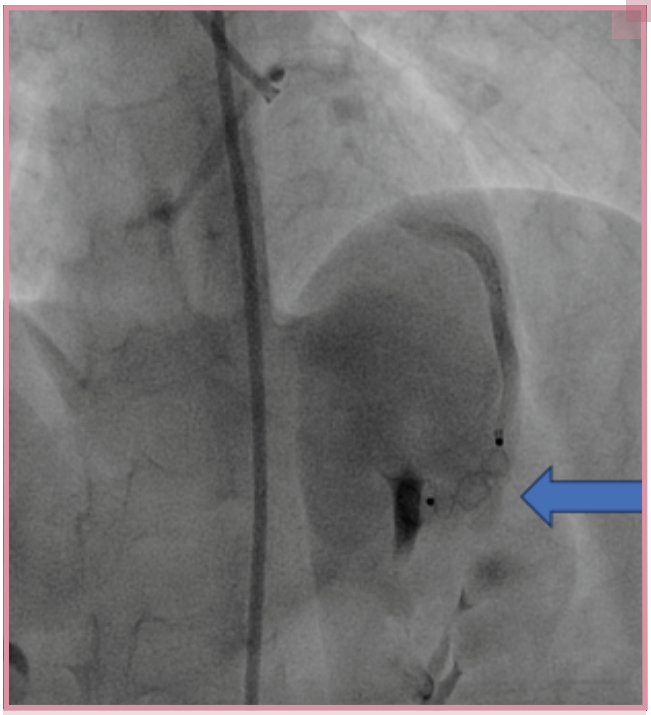

Figure 3 : Distal LAD occluded by Vascular Plug with no filling of RV

Discussion

Coronary artery fistula (or coronary-cameral fistula) in the majority of cases (>90% of patients), the coronary fistula results from an abnormal coronary artery system with aberrant termination rather than true arteriovenous fistula. The right coronary is most commonly involved in coronary artery fistula. In more than 90% of reported cases, the fistula terminates in the right side of the heart (either to the RV or the PA; less commonly to the RA). It rarely terminates in the left side of the heart, but when it does, the majority enters the LA. The pathophysiological importance of a coronary fistula is related to the volume of blood flowing and the pressure gradient through communication.[2]

Patients usually are asymptomatic. However, CHF may develop if the shunt through the fistula is large. With a significant shunt, a continuous murmur, similar to the murmur of PDA, is audible over the precordium (rather than in the left infraclavicular area).

Echocardiographic studies usually suggest the sites and types of the fistulas. The presence of a massively dilated proximal portion of one coronary artery while the other coronary artery is of normal size suggests coronary artery or arteriovenous fistula. The dilatation is usually uniform. Often selective coronary artery angiography is necessary for an accurate diagnosis before the intended intervention.

Now a days CT coronary angiogram also done to profile Fistula in better 3D dimension which help in choosing the proper implant (Vascular Plugs or Coils) and sizes.

Most children with this condition are asymptomatic. Spontaneous closure may occur in small fistulae. However, some patients may present with symptoms of dyspnoea on exertion, increased fatigability, and possibly signs of high-output CHF. Rarely, adult patients may present with angina, palpitation, or signs of exercise-related coronary insufficiency.

Fistula-related complications are not common; reports of aneurysmal formation, infective endocarditis, pericardial effusion, and supraventricular and ventricular arrhythmias have been associated with coronary-cameral fistulae infrequently.[3]

Small fistulous connections in the asymptomatic patient may be monitored. For a moderate or large coronary artery fistula, transcatheter occlusion is reasonable using coils or other occluding devices. Elective surgery is indicated if not amenable to catheter occlusion. Using cardiopulmonary bypass, the fistula is ligated as proximal as can be done without jeopardizing flow in the normal arteries, and it is ligated near its entrance to the cardiac chamber[4]

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Acute Hyperkalaemia in an Infant with Acute Gastroenteritis with Severe Dehydration with Pre-Renal AKI

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Introduction

Acute gastroenteritis (AGE) is a major cause of morbidity and mortality in children under the age of five, with 3-5 billion cases and approximately 2 million fatalities happening each year, primarily in the resource-limited world [(1).] The most important and dangerous complications related to AGE are dehydration, metabolic acidosis, and electrolyte disturbances. Kidney hypoperfusion secondary to dehydration is one of the most common causes of paediatric acute kidney injury (AKI), contributing to 12% of AKI in children [(2).] Children in hospitals who develop AKI could have longer hospital stays, higher mortality, and a longer risk of developing chronic kidney disease (CKD), hypertension, and proteinuria [(3,4).] Here, we describe a case of prerenal AKI due to Acute gastroenteritis with severe dehydration.

Report of Case

A two-month-old male baby, born to a non-consanguineously married couple was brought to the paediatric emergency by his parents with complaints of fever, vomiting, and loose stools for three days. The baby was lethargic, was difficult for him to wake up, and had not passed urine throughout the day. There was no significant medical history. On admission, his blood pressure was 90/60 mmHg; pulse was 135 beats per minute; and the temperature was 37.8 °C. His body weight was 4.3 kg, 8.5% less than his pre-illness weight of 4.7 kg. On examination, the child had depressed anterior fontanelle, sunken eyes, and delayed skin pinch, all suggestive of severe dehydration. Peripheral IV catheter was inserted and blood was drawn for investigations and fluid bolus NS @10 ml per kg was given. Lab reports revealed, Sodium 131 mmol/L, Potassium- 9.6 mmol/L, Urea-131mg/dl, and Creatinine-2.3 mg/dl. ABG was s/o Severe metabolic and respiratory acidosis, with pH 7.08, PCo2-29.4, HCO3-8.6, BE- -19.9. I/v/o severe Hyperkalaemia, IV calcium gluconate was given under cardiac monitoring, along with Salbutamol nebulisation, and K-Bind sachets through an NG tube. Plan C of dehydration correction was given. Ultrasound KUB s/o left minimal hydronephrosis with an RPD of 6mm.2D ECHO was done to rule out heart failure/myocarditis-results were within normal limit. Initial investigations revealed Neutrophilic leucocytosis with raised CRP. Hence IV ceftriaxone was started after sending blood culture. Antibiotics were stopped once the blood culture showed no growth. Serum NT PRO BNP levels done - 1094 pg/ml. After 24 hours of fluid therapy, the child showed gradual improvement with Potassium in decreasing trend reaching a final value of 4.5 mmol/l. Repeat RFT and ABG was normal. Refractory hyperkalaemia with increase in positive balance and increase in oxygen requirements are indication for dialysis. Dialysis was planned if adequate response was not achieved with our line of management. As the child responded well he was discharged with no residual symptoms.

Day of Hospitalisation	Serum Potassium Levels (mmol/l)	Serum Creatinine Levels (mg/dl)
DAY 1	9.6	2.32
DAY 2	5.92	1.61
	3.99	
DAY 3	3.16	0.95
DAY 4	4.58	0.30

Discussion

Acute gastroenteritis typically resolves on its own but in more severe cases in babies and children, where symptoms including fever, vomiting, and severe diarrhoea can gravely exacerbate dehydration, requiring intensive care. Most cases of acute diarrhoea in developing countries are caused by infectious

gastroenteritis; patients may present with watery diarrhoea or invasive (bloody) diarrhoea (5). Rotavirus, Norovirus, and Adenovirus are the most common causes of infantile diarrhoea (6). Fluid and electrolyte replacement are the mainstay of treatment to prevent and correct severe dehydration and acidosis (7). Several variables can contribute to hyperkalaemia in AKI patients (8). These include decreased glomerular filtration rate (GFR), decreased tubular potassium secretion, tissue breakdown with intracellular potassium release, and metabolic acidosis with transcellular potassium movement (each 0.1 unit decrease in arterial pH raises serum potassium by 0.3 mEq/L). The urgency of treatment of hyperkalaemia varies with the level of extracellular (serum/plasma) potassium, the rapidity of the increase in potassium, and the presence of associated hyperkaliaemic symptoms (9). Severe hyperkalaemia (potassium level □7 mEq/L [mmol/L]) is a serious medical problem that requires immediate attention. Acute rapid transient measures include intravenous (IV) calcium infusion to stabilize the cardiac membrane and followed by interventions that shift potassium from the extracellular space into the cells, including Inhaled beta-adrenergic agonists, IV Insulin, and Glucose, IV Sodium bicarbonate for correction of acidosis. Increasing the extracellular pH with sodium bicarbonate leads to hydrogen ion movement from the cell into the extracellular space. As a result, extracellular potassium moves into the cell to maintain electroneutrality, hence correcting the acidosis helps in reducing the potassium levels. Strong suspicion of underlying bacterial infection along with an increase in infection parameters in the laboratory results led to the administration of ceftriaxone which was discontinued 3 days later due to negative bacterial culture results. The diarrhoea and dyselectrolytemia improved after fluid therapy and the baby was discharged with normal vitals.

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Abdominal Angioedema: A Surgical Abdomen Mimic

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Introduction

Angioedema is a condition characterized by localized, transient swelling of the deeper layers of the skin or mucous membranes in the upper respiratory or gastrointestinal tract. This swelling tends to occur in areas with loosely attached skin and can manifest as acute severe abdominal pain, which is unresponsive to usual analgesics.

Case Details

We present the case of a 9-year-old boy who presented with acute severe abdominal pain, accompanied by fever, urticarial rash, and loose stools. He had been on oral antibiotics and steroids for two days. Physical examination revealed Wt-50kg, Temp. 99*, PR-114/min, SpO2-98% (room air), BP- 110/70mmHg, GCS-14/14 with mild hepatosplenomegaly and tenderness in the right iliac fossa.

Investigations

Test	Result
Hb/	14.1/ 42.5 - 11.3
TLC/DLC	14.28/ N78 L20 E1 M1 11.59
PBF	no immature cells or toxic granules, platelets adequate
CRP	113.2 mg/dl 97 (day 3) 3.4 (day 10)
Procalcitonin	0.412 ng/ml
IgE	462.5 IU/ml
Stool OB/ routine	Negative
Urine	Protein +, WBC- 2-3, RBC-nil
Serum Tryptase	23ng/ml (<11) 10 (day 60)
C1q complement	5mg/dl
C3, C4	normal
Typhidot IgM	Negative
CECT Abdomen	Hepatosplenomegaly (L-17cm, S-12.3cm). Circumferential thickening of terminal ileum for a length of 6cm, few discrete subcentimetric mesenteric lymph nodes - ?infective
Urinary Histamine (24hr)	0.044 mg/24hr (0.006-0.131)
Skin Prick Test (after 6wks)	Positive reactions to Dust mite and Mugwort

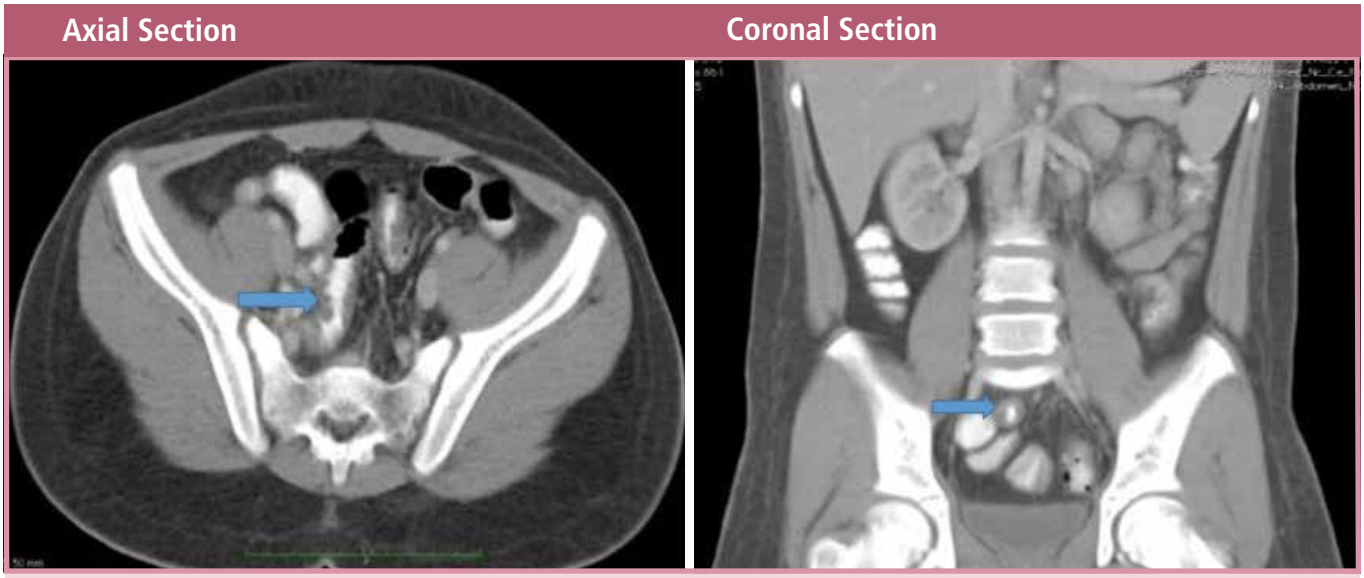

Figure 1 : Intestinal wall thickness with contrast filled bowel on CECT Abdomen

Final Diagnosis

Acute Allergic Abdominal Angioedema

Discussion

Angioedema occurs due to a rapid increase in permeability of submucosal or subcutaneous capillaries and post-capillary venules, resulting in localized plasma extravasation. This process involves the release of vasoactive substances such as histamine or bradykinin. Angioedema can be classified into four types: acute allergic angioedema, non-allergic drug reaction, idiopathic/chronic angioedema, and hereditary angioedema. The duration of angioedema attacks typically ranges from 1 to 5 days, depending on the underlying cause.

The effects of visceral angioedema can vary, ranging from life-threatening episodes to varying degrees of abdominal pain. Radiographic imaging may reveal transient findings such as obstruction, ascites, and oedematous viscera. Laboratory tests are generally not helpful, but diagnostic markers such as serum histamine and tryptase levels can aid in confirming the diagnosis. It is estimated that 10% to 20% of people worldwide will experience an episode of angioedema or urticaria at some point in their lifetime, with women being more prone than men.

Conclusion

Isolated abdominal pain due to angioedema, without associated skin or respiratory symptoms, can lead to incorrect diagnoses or unnecessary interventions. Cases of acute abdomen that do not respond to usual analgesics should be evaluated with caution to consider the possibility of angioedema as the underlying cause.

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Multiple Cardiac Rhabdomyomas in Dizygotic Twins

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Keywords : Rhabdomyomas, Dizygotic, Twins, Tuberous sclerosis complex, Cardio-oncology

Abstract

Multiple cardiac rhabdomyomas are usually found in association with the tuberous sclerosis complex (TSC). We report a case of multiple cardiac rhabdomyomas in a set of dizygotic twins with no other signs and symptoms of TSC in the twins or parents.

A 26-year-old woman with dizygotic twin pregnancy underwent fetal echocardiography in the third trimester, at 34 weeks of gestation. A rounded echogenic lesion was noted in the right ventricle of one of the fetuses (maternal left side) measuring 13 × 12 mm. The second fetus appeared normal. Subsequent pregnancy remained uneventful. The twins were delivered at term (37+3 weeks) via cesarean section. Twin 1 weighed 2.66 kg at birth and had Apgar scores of 8 and 9 at 1 minute and 5 minutes, respectively. Twin 2 weighed 2.52 kg and had Apgar scores of 8 and 9 at 1 minute and 5 minutes, respectively. The twins were referred to us, and postnatal echocardiography was performed on day 3 post-delivery. Echocardiography in twin 1 revealed multiple echogenic masses of varying sizes in the right ventricle and left ventricle, consistent with a diagnosis of multiple cardiac rhabdomyomas (Figure 1). The largest mass was located in the right ventricular outflow tract (RVOT) and measured 13 × 10 mm. It led to mild RVOT obstruction, with a pressure gradient of 14 mm Hg across the RVOT. A patent foramen ovale with bidirectional shunt was also noted. The remainder of the examination was normal.

Echocardiography in twin 2 also revealed 3 small echogenic masses, consistent with a diagnosis of multiple cardiac rhabdomyomas. All 3 masses were intramural. One was located at the base of the interventricular septum, one in the lateral wall of the right ventricle, and one in the apex of the right ventricle (Figure 2). A patent foramen ovale with right-to-left shunt was also seen. Physical examination revealed no cutaneous manifestations of TSC. There were no episodes of seizures since birth. Neither parent had any manifestation of TSC on history and physical examination. No family history of TSC was present.

Further workup and genetic testing for TSC was planned. Twin 1 was planned for everolimus therapy in view of the large tumor obstructing the RVOT. However, the

parents refused, and the twins were lost to follow-up.

We found 3 other case reports of cardiac rhabdomyomas in twins, highlighting the rarity of this case presentation. Of these, 2 cases described multiple cardiac rhabdomyomas in monozygotic twins. Only 1 case involved multiple cardiac rhabdomyomas in dizygotic twins. Intrauterine demise of 1 of the twins occurred at 24 weeks gestation. Similar to our case, the parents in this case were clinically unaffected. The surviving twin and father were subsequently found to carry 2 TSC2 mutations on molecular testing. Our case differs in that both fetuses survived the pregnancy and were born well.

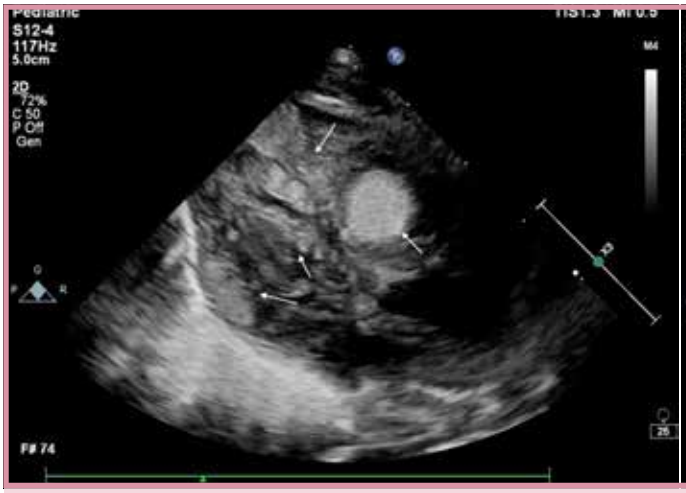

Figure 1 : Twin 1: two-dimensional transthoracic echocardiography parasternal long-axis view, systolic phase, demonstrates multiple well-defined, hyperechoic homogenous masses of varying sizes in both the right and left ventricles (arrows)

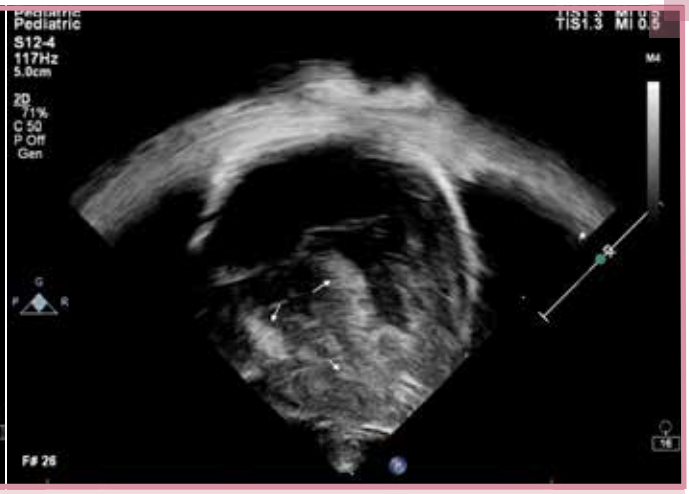

Figure 1 : Twin 2: two-dimensional transthoracic echocardiography apical 4-chamber view demonstrates intramural rhabdomyomas embedded in the myocardium at the base of the interventricular septum, lateral wall, and apex of the right ventricle (arrows)

Colorful tribute to **Dr Arvind Kumar** by his little Patient

Congenital CMV Infection - A Case Report

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Introduction

"A late preterm, 36+4 weeks male baby weighing 2.83 kg was born by caesarean section, with the indication being fetal ascites, hepatosplenomegaly, and MCA redistribution pattern with an alteration of cerebroplacental ratio. The baby cried immediately after birth with good APGAR scores. The new-born was admitted to the NICU due to prematurity, ascites, and respiratory distress. The baby was started on High-flow nasal cannula (HFNC) support. The chest X-ray showed no evidence of Respiratory Distress Syndrome, but bilateral streaky and patchy opacities were seen. The baby had significant hepatosplenomegaly, with the liver measuring 5 cm and the spleen measuring 3 cm, along with ascites. No limb or chest wall edema was noted. On the first day, the baby developed purpuric spots over the abdomen. Due to the significant hepatosplenomegaly, ascites, and the presence of petechiae and purpuric spots, congenital CMV infection was considered and investigated. Laboratory data showed thrombocytopenia (platelet count of 70,000/µL), ALAT (51 U/L), ASAT (168 U/L), total bilirubin (9.87 mg/dL), conjugated hyperbilirubinemia with direct bilirubin (5.09 mg/dL), alkaline phosphatase (ALP) of 139 U/L, and gamma-glutamyl transferase (GGT) of 89 U/L. The activated partial thromboplastin time (aPTT), prothrombin time (PT), and international normalized ratio (INR) were mildly elevated. Postnatal abdominal ultrasound confirmed hepatosplenomegaly and ascites, while the cranial ultrasound revealed grade 1 intraventricular hemorrhage (IVH) with mild ventricular enlargement. The CMV IgG was 212.90 AU/mL (high), CMV IgM was 1.22 (reactive), and CMV avidity was 48%. CMV DNA PCR was found to be negative in the peripheral blood and urine samples. The maternal CMV IgG was 226.90 AU/mL. Intravenous ganciclovir administration (12 mg/kg/day) was started, and platelet transfusions were given for thrombocytopenia. After 2 weeks of treatment, there was clinical improvement with no further oxygen requirement, and the platelet counts increased to a normal level (165,000/µL). After 2 weeks of intravenous treatment, there was further improvement, with platelet counts increasing to 329,000/µL and no petechiae observed. The latest liver parameters were as follows: direct bilirubin at 6.7 mg/dL, ALAT at 104 U/L, and ASAT at 274 U/L. The repeat CMV IgG was 605.20 AU/mL. The newborn is being subsequently followed up, growing well, and we have switched to an oral antiviral drug (valganciclovir 32 mg/kg/day) which the baby will require for four weeks. Eye screening revealed no evidence of chorioretinitis, and a hearing assessment (BAER) is being planned."

Discussion

CMV infection is the most common congenital viral infection, with prevalence ranging from 0.2% to 2.2% in different parts of the world [1]. Congenital CMV infection is the leading infectious cause of congenital malformations, resulting in sensorineural hearing loss, vision impairment, and cognitive deficits in childhood [2]. Vertical transmission of CMV to the foetus can occur during the intrauterine, perinatal, or postnatal period, including through exposure to breast milk. However, diagnosing maternal primary cytomegalovirus infection can be challenging clinically, as 25-50% of cases are asymptomatic and the symptoms are often non-specific [3]. Maternal seroconversion, determined by the detection of CMV IgG antibodies, is considered the gold standard serological investigation. In our case, the CMV IgG antibody was positive, with a low avidity of IgG antibodies to CMV and an increase in CMV IgG titre.

Neonates with congenital cytomegalovirus infection can be either asymptomatic or symptomatic (10%) at birth. There is a predilection for involvement of the central nervous system and reticuloendothelial system. Even asymptomatic newborns can experience long-term sequelae, such as sensorineural hearing loss. The most

frequent clinical signs observed include hepatosplenomegaly (60%), microcephaly (53%), jaundice (67%), petechiae (76%), and at least one neurological abnormality (68%) (Reference 4). In our case, the newborn presented with prematurity at 36+4 weeks, jaundice, petechiae on the trunk and limbs, hepatosplenomegaly, and ascites. The frequency of biological abnormalities in congenital CMV infection includes transaminitis (83%), thrombocytopenia (77%), and hyperbilirubinemia (69%) (Reference 4). Our case exhibited transaminitis, thrombocytopenia, hyperbilirubinemia, and cholestasis. Additionally, the baby had Grade 1 intraventricular hemorrhage (IVH) with ventriculomegaly on initial scans. The latest ultrasound showed resolving hematoma in the left caudothalamic groove with a prominent frontal horn of the left ventricle and no calcifications noted. The mortality rate among symptomatic newborns ranges from 5% to 10%. Long-term complications may include psychomotor retardation (70%), sensorineural hearing loss (50%), and optic atrophy or chorioretinitis (20%).

Confirming a positive cytomegalovirus screening result should involve testing a subsequent sample (either saliva or urine) collected within the first 3 weeks of life. However, in our case, urine CMV PCR yielded a negative result.

Intravenous ganciclovir and oral valganciclovir are the current medications available and studied for the treatment of infants with congenital cytomegalovirus infection (Reference 5). In our case, we administered intravenous ganciclovir at a dosage of 12 mg/kg/day for 2 weeks, followed by oral valganciclovir at a dosage of 32 mg/kg/day, which we plan to continue for 4 weeks. The new-born is being regularly monitored and followed up.

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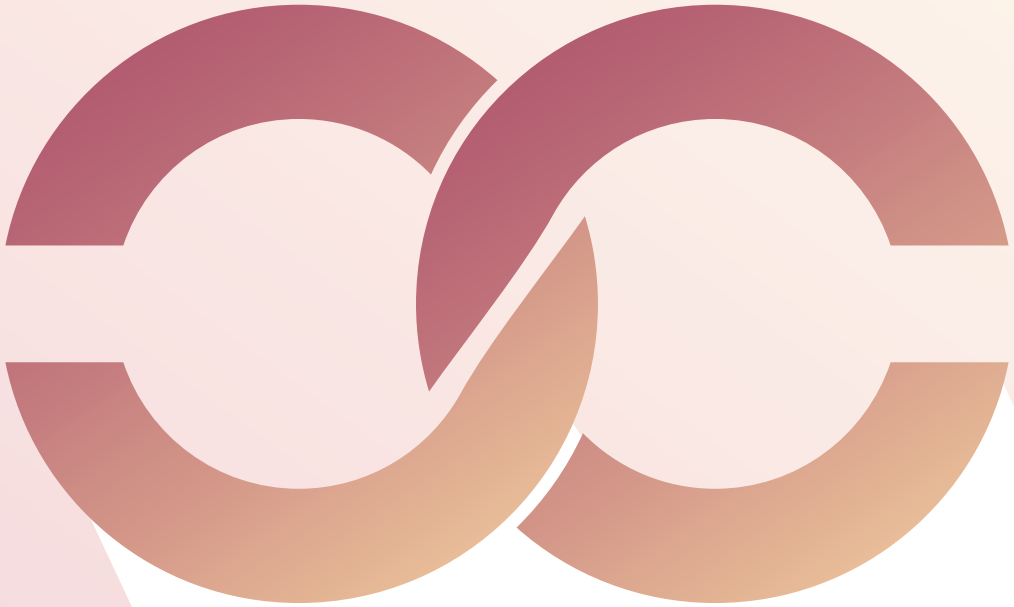

PAEDIATRIC NUTRITION AND DIETETICS

Diet in Laryngeal Cleft

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Laryngeal Cleft

A laryngeal cleft (or laryngotracheal cleft) is an abnormal opening between the larynx and the esophagus through which food and liquid can pass through the larynx into the lungs. This causes a number of eating and breathing problems. Laryngeal cleft is congenital problem.

Signs and Symptoms

A child having a laryngeal cleft can have a wide range of signs and symptoms such as:

- Coughing
- Choking with feedings
- Shortness of breath
- Poor weight gain
- Hoarseness
- Apnea
- Stridor
- Frequent pneumonia
- Aspiration
- Gastric esophageal reflux

Laryngeal clefts are classified in one of four ways:

- **Type I** is the mildest form of laryngeal cleft. The gap between the larynx and the esophagus is located above the vocal cords.
- **Type II** laryngeal cleft extends into the lower cartilage of the voice box, below the vocal chords.
- **Type III** laryngeal cleft extends beyond the voice box and into the trachea.
- **Type IV** is the most severe form laryngeal cleft. The gap extends even further down into the windpipe, and may go all the way to the bottom of the trachea.

Cleft severity and comorbidities contribute to symptom severity. Low severity, mildly symptomatic laryngeal clefts may be managed conservatively with feeding modifications, while more severe clefts require surgical intervention. Dysphagia persisting despite medical optimization and feeding modifications is a major indication for surgical intervention.

Diet for Child with Laryngeal Cleft

With laryngeal cleft, there is a risk for aspiration. A dysphagia diet can help prevent aspiration. It has different levels based on the thickness of the food. The levels can be increased gradually based on the condition and acceptability of the patient.

A dysphagia diet ranges from stage 0 to 7. Zero indicates thin liquids (0), followed by slightly thick (1), mildly thick (2), moderately thick liquid (3). Beyond this stage is the solids that is presented in different textures-pureed (4), minced and moist (5) soft and bite sized (6), easy to chew and lastly regular food (7).

For patients with risk of aspiration, thin liquids should be avoided and the feeds can be started from pureed semi solids(stage 4) This can be advanced to minced, soft and finally regular normal diet based on the tolerance. The following description explains the stages with examples

Stage 4 - Pureed Food

These foods:

- Can often be eaten with a spoon, but sometimes a fork
- Can't be drunk from a cup
- Don't need to be chewed
- Are not sticky or lumpy
- Fall off a spoon all together when tilted and still hold shape on a plate.
- Can't be poured but move very slowly if the plate is tilted
- For example, mashed khichdi or mashed soft rice and dhal, pudding, thick kheer, broken wheat porridge etc

Stage 5 - Minced, Moist

- Can be eaten with a fork or spoon
- Can be scooped and shaped on a plate.
- Are soft and moist but don't separate into liquid
- May have small lumps that can be mashed with the tongue

For example, spoon mashed soft rice with dhal and vegetables, boiled and spoon mashed carrot, bread well mixed in milk, spoon mashed banana/papaya

Stage 6 - Soft

These foods:

- Are tender, moist, and bite-sized
- Can be eaten with a fork or spoon
- Must be chewed
- For example boiled egg, wheat dosa, Neer dosa, Besan ka cheela, soft idli, omelette

Stage 7 - Regular

- Normal, everyday foods of varying textures, including soft, stringy, and hard and crunchy
- Foods that can be eaten by any method. For example, from a cup or using utensils.
- Foods that need to be chewed, with all types of textures and may have pieces that can't be swallowed,
- For example, Chapathi, toasted bread etc

Preparing Food and Liquids

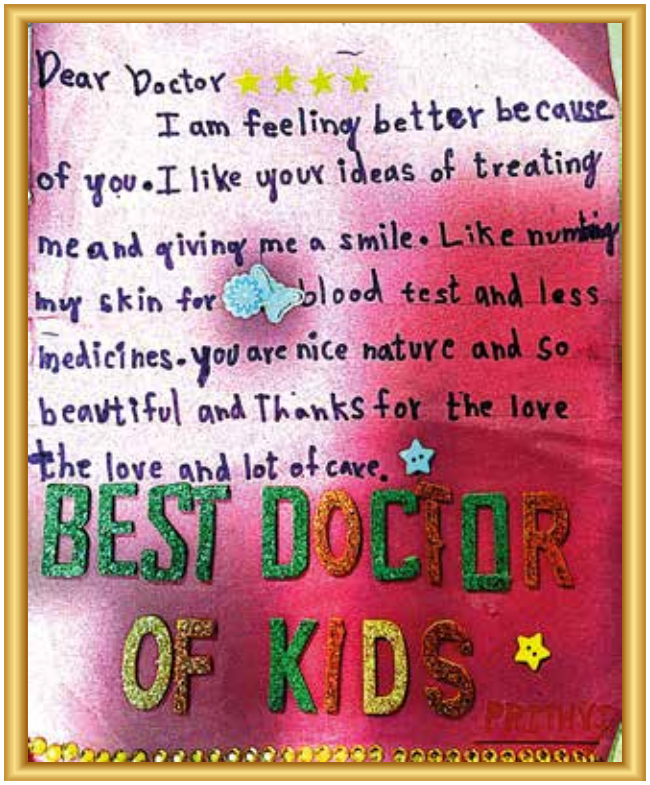
The following steps may help in easing up the intake of food.

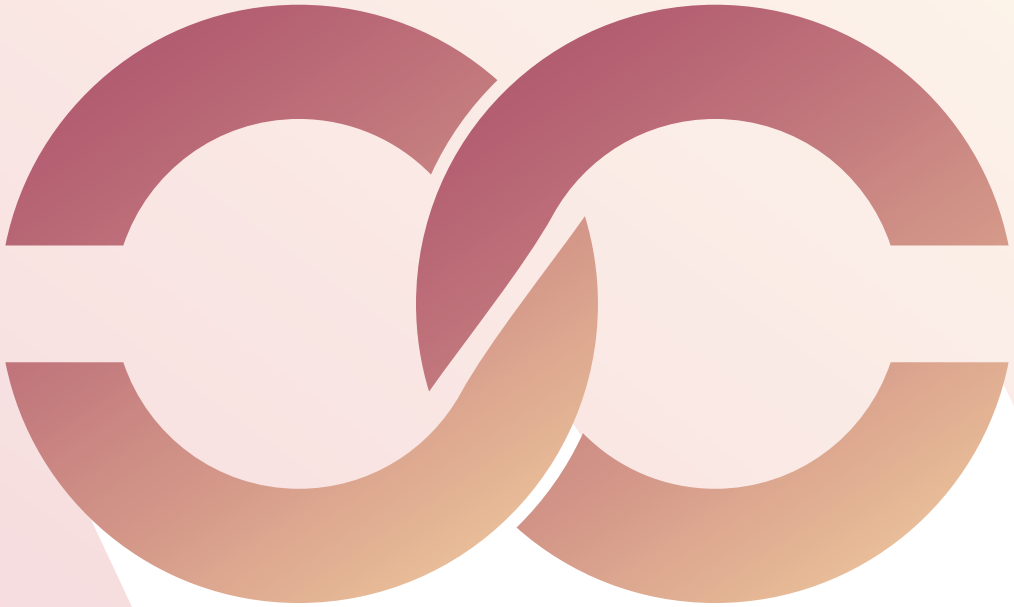
- Thin liquids like coffee, tea, milk, buttermilk, juice, milkshake or soup should be avoided as they increase the risk of aspiration.
- For thickening the food,thickening agents can be added to the liquids wherever required. For example, corn starch, Maida(multipurpose flour), fine semolina(chiroti rava),rice flour,Besan(roasted Bengal gram flour) can be added to thicken soups,gravies etc.Milk powder or custard powder can be added for baby foods.
- Bland unpalatable food should be avoided. Food can be seasoned and spiced and then pureed to increase the acceptability by the patient.

Precautions to be taken while feeding the patient

- While eating or drinking, it may help to sit upright, with back of the patient straight.
- Infants must be burped often.
- Support pillows can be used to get into the best position.
- It may also help to have few distractions while eating or drinking
- The patient must stay upright for at least 30 mins after having food to avoid the reflux.

Thus, the nutritional status of the patient with laryngeal cleft can be improved by modifying the texture of feeds, individualizing the diet based on the condition (pre or post repair) and tolerance.





COVID-19

Multisystemic Inflammatory Syndrome in Children (MIS-C)-A Post Covid Phenomenon

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Multisystem inflammatory syndrome in children is a serious disorder associated with Covid 19 infection. Essentially, it is an immune mediated response to the Covid virus and may occur during the active Covid infection or a few days to weeks after Covid.

As the pandemic started in early 2020 and adults started suffering from severe Covid, many children started showing up with syndrome of fever associated with involvement of multiple systems of the body without evidence of any other infection. Soon the paediatricians in China and Europe realized that this is a syndrome associated with Covid and is being seen only in children. WHO quickly came out with diagnostic criteria [(1)] (Figure 1), followed by the CDC and IAP.

A few months later, when the first wave of Covid came to India and also in the second wave, large number of such patients were identified here too. All the tertiary care hospitals started receiving these critically ill children. Many of them had cardiac involvement (including dilatation of coronary arteries), shock and even CNS involvement. However, by now it had been realized that this is a hyper immune response and can be controlled with steroids and iv immunoglobulins. Early recognition and appropriate treatment reduced mortality and morbidity significantly.

In a multi centric study, several hospitals of the state of Haryana, including FMRI, pooled their data and analyzed it from the viewpoint of severity of the disease and cardiac involvement [(2).] Disease severity was graded (mild/ moderate/severe) and presence of cardiac abnormalities noted. Patients with and without cardiac abnormalities and with and without severe disease were compared.

Forty-eight children with MIS-C were included (median age - 9.5 y). Fever (100%), gastrointestinal (83.3%) and mucocutaneous (50%) symptoms were common. Only 16.7% of patients had a previous history of documented SARS-CoV-2 infection. Severe disease and cardiac abnormalities were seen in 47.9% and 54.2% patients, respectively. NT- proBNP > 1286.5 pg/mL and thrombocytopenia (□119500/μL) were significant risk factors for severe MIS-C. Forty-five patients (93.8%) recovered and 3 died. The median hospitalization duration was 7 d (5–9.5).

MIS-C is a newly recognized entity and closely mimics the earlier well-known disorders like Kawasaki disease, Septic shock and Hemophagocytic Lympho- Histiocytosis (HLH). Release of diagnostic criteria by WHO early in the pandemic and close online collaboration among all the stake holders helped in saving many lives from this devastating illness in that era of restrictions and lock outs.

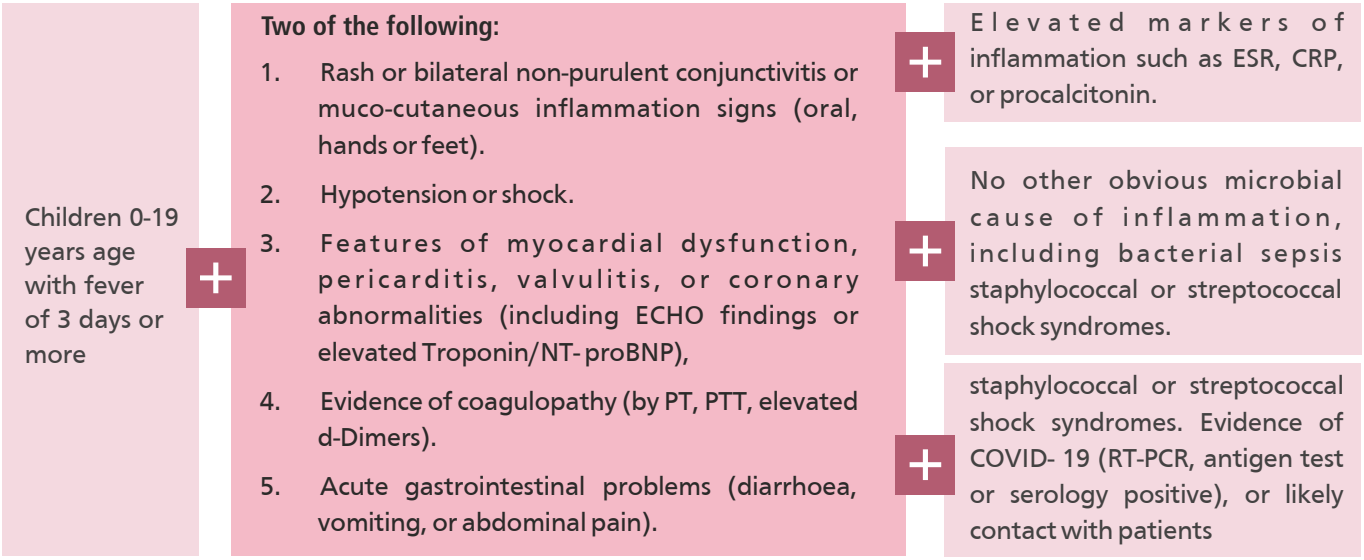

Figure 1 : Multisystemic Inflammatory Syndrome in Children (MIS-C); WHO Definition

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Neonatal MIS-C - Managing the Cytokine Storm

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Abstract

A term baby girl with uneventful ante-natal history had an erythematous rash followed by fever from day eight. She was diagnosed to have late onset sepsis and treated accordingly. She received immunoglobulin for persistent thrombocytopenia following which there was a transient improvement. Baby was transferred to our hospital on day twenty-five, following recurrence of fever, watery diarrhea and a generalized maculopapular rash. On admission, she had tachycardia, tachypnoea, anemia, thrombocytopenia, hypoalbuminemia and generalized edema. RT-PCR for COVID-19 was positive. Within twelve hours of admission, baby developed cardiogenic shock with pulmonary oedema and needed invasive ventilation. Echocardiography revealed ejection fraction of 40% with mild pericardial effusion. NT-Pro BNP was 33000gm/L, D-dimer 16500 µg/L and ferritin 16000 ng/ml. Methylprednisolone, immunoglobulin and enoxaparin was started with a diagnosis of multisystem inflammatory syndrome associated with COVID-19 (MIS-C). She developed seizures, pulmonary hemorrhage and cardiac arrest the following day along with acute kidney injury. She was extubated after five days. Steroid was stopped after five days as she developed hypertension and echocardiography had normalized. Five days after extubation she again developed respiratory distress and was ventilated again for two days. Echocardiography showed moderate LV-dysfunction along with secondary elevation of ferritin. Methyl-prednisolone was restarted and continued for five days followed by tapering dose of oral prednisolone on which she was finally discharged. Though mild myocarditis with COVID-19 has been reported, MIS-C in a newborn with refractory myocarditis along with gastrointestinal and renal manifestations is a rare entity. Dermatological manifestation of neonatal COVID-19 is also unique.

Case Report

A term baby girl with uneventful ante-natal history was noted to be febrile on day eight with an erythematous, generalised, fleeting rash with facial sparing (pic 1a). As fever persisted, the baby was hospitalised on day 10 where she was provisionally diagnosed as late onset sepsis and started on Meropenem and Amikacin. However, noting a progressive rise in C Reactive protein with thrombocytopenia [Table 1] on day twelve she was referred to another hospital. On second admission, sepsis screen was repeated [Table 1] and blood culture grew coagulase negative Staphylococcus aureus. Meropenem was continued and Teicoplanin was added. Within 48 hours (day seventeen), she was noted to be afebrile and rashes had subsided. She received intravenous immunoglobulin (IVIG) at 1g/kg after two successive days of platelet transfusions as platelet count persistently remained less than 8x109/micro L. This was followed by a progressive rise in platelet count. Echocardiography was normal. Ultrasonography of abdomen showed hepatomegaly with minimal ascites. She had clinical improvement and was on oral feeds by day seventeen.

On day twenty-four, fever recurred with rise in sepsis markers [Table 1]. On day twenty-five, acute onset disseminated erythematous maculo papular skin lesions were noted [Figure 1b, 1c, 1d]. There was sparing of the face with involvement of the neck, elbow, knees and a necrotic lesion in left groin. Repeat blood culture did not

grow any organism. The baby was shifted to our hospital on day twenty-five.

On admission, she was febrile, pale and tachycardic (heart rate 180-200/minute) with hepatosplenomegaly and greenish watery stool. She was hemodynamically stable and on minimal oxygen of 2litres/min could maintain saturation of 95%. Antibiotics were changed to Cefoperazone-Sulbactum, Flucloxacillin and Clindamycin considering a differential diagnosis of staphylococcal or pseudomonal sepsis. Reverse transcriptase –polymerase chain reaction (RT-PCR) for COVID-19, which was done routinely as hospital protocol in view of the COVID pandemic, was positive on admission. Overnight there was rapid deterioration with progressively increasing respiratory distress and mixed respiratory and metabolic acidosis in arterial blood gas for which baby needed invasive ventilation. Chest X-ray showed pulmonary edema and cardiomegaly. Echocardiography showed significant systolic dysfunction with ejection fraction of 40% and mild pericardial effusion. Adrenaline infusion was started and continued for three days. With high grade fever, and multisystemic involvement (respiratory involvement needing ventilation, cardiac involvement, dermatological involvement in the form of rash, GI involvement in the form of diarrhoea) as well as high inflammatory markers like elevated CRP, ferritin, N-terminal pro B-type natriuretic peptide (NT –pro BNP) and D-Dimer, a diagnosis of multisystem inflammatory syndrome associated with COVID-19 (MIS-C) was suspected. She was initiated on IVIG 2g/kg over 24 hours along with methylprednisolone at 2mg/kg/day. Enoxaparin was also started at therapeutic dose (1mg/kg/dose twice daily) which was subsequently changed to prophylactic dose (1mg/kg/dose once daily) as D-dimer reduced to <1500µg/L after seven days. We did consider but finally decided not to use any antiviral.

On day twenty-seven, she had a short duration seizure which was controlled with Phenobarbitone. Lumbar puncture was not done as baby was too unstable and EEG was not done as seizures never recurred and Phenobarbitone was stopped after five days. An MRI scan on day forty-three revealed no abnormality. On the same day, i.e. day twenty-seven, she also had pulmonary haemorrhage and a cardiac arrest and was resuscitated as per neonatal guidelines. Post resuscitation she developed acute kidney injury with oliguria (Urine output 0.7 ml/kg/hr) and deranged renal function (Serum creatinine - 1.9mg/dl). Ultrasonography of kidneys was suggestive of renal parenchymal disease with normal doppler flow in the renal vessels. She was conservatively managed with albumin (Serum albumin was 1.8mg/dl) and furosemide. Anemia (Haemoglobin 6.7mg/dl) was corrected with packed red blood cell transfusion. The baby was finally extubated to heated humidified high flow nasal cannula (HHHFNC) after five days and feeds were initiated. Bronchoalveolar lavage grew Klebsiella and antibiotics were changed to Tigecycline and Colistin in renal adjusted dose as per sensitivity reports. High resolution computed tomography (HRCT) of the thorax showed atelectasis of both lower lobes of lung. Repeat COVID-19 RT-PCR was negative at seven days. Repeat echocardiography at day thirty suggested normal cardiac function with ejection fraction of 64%. Steroids were stopped after five days as baby developed hypertension which was controlled with amlodipine and propranolol.

She developed feeding intolerance followed by a gradual deterioration of her respiratory status 5 days after extubation (Day thirty-five) and had to be re-ventilated following a failed CPAP trial. Repeat echocardiography showed moderate left ventricular (LV) systolic dysfunction, generalised LV wall hypokinesia with ejection fraction of 35- 40%. She had a second rise in ferritin [Table 1]. Milrinone was administered and methylprednisolone was restarted. Baby showed clinical improvement over the next two days and was extubated to HHHFNC on day thirty-seven of life. She was febrile yet again. Bronchoalveolar lavage grew Klebsiella which was now sensitive to Meropenem which she received for two weeks. Feeding was re-established and sepsis markers and renal function improved over the next few days. Intra venous methyl prednisolone was given for five days followed by oral prednisolone. She was eventually discharged on day fifty of life on tapering dose of prednisolone, subcutaneous

TABLE 1 with amlodipine and propranolol.Important Laboratory Parameters and Echocardiogram Finding

Blood	Day 12	Day 13	Day 14	Day 17	Day 24	Day 25	Day 26	Day 28	Day 33	Day 35	Day 42
Hb, mg/dL	10.8	9.0	8.2	8.5	7.8	6.5	12.5	11.5	10.1	7.7	13.5
TLC, mm^3	23 10^3	15 10^3	14 10^3	22 10^3	7 10^3	12 10^3	18.8 10^3	17 10^3	8 10^3	18 10^3	15 10^3
DC	N78, L22	N80, L18	N71, L21	N70, L28	N43, L55	N66, L23	N72, L20	N40, L50	N58, L36	N68, L25	N75, L18
PLT, mm^3	105 10^3	10 10^3	5 10^3	110 10^3	90 10^3	100 10^3	130 10^3	150 10^3	80 10^3	135 10^3	135 10^3
CRP, mg/L	28	78.5	37.7	9.5	44	29	68	49	63.9	99.8	13
Ferritin, µg/L	—	550	—	—	—	16 500	—	1702	1265	1911	—
NT-ProBNP, pg/mL	—	—	—	—	—	—	33000	—	—	11 900	—
D-dimer, ng/mL	—	—	—	—	—	16 500	—	1702	—	1265	1140
Echocardiography	—	—	Normal	—	—	Systolic dysfunction, EF 40%	—	Good LV function, EF 64%	—	LV dysfunction, EF 35%	Good LV function, EF 60%

DC, differential count; EF, ejection Fraction; Hb, hemoglobin; PLT, platelet count; TLC, total leukocyte count; —, not applicable.

low molecular weight heparin (prophylactic dose) and vitamin supplements. Baby is well on follow up with no further recurrence of any clinical signs of illness and normal echocardiogram with good left ventricular function and 65% ejection fraction.

Discussion

The World Health Organization described COVID-19 as a public health emergency on 31st January 2020 [1]. Severe acute respiratory syndrome coronavirus type 2 (SARS-CoV-2) has affected all age groups. However, there are limited case reports in the neonatal population [2-4]. They are usually asymptomatic or present with subtle clinical manifestations. There are sporadic case reports of neonates being positive for SARS-CoV-2 within 48 hours of birth. However, vertical transmission is still not an established entity. In April 2020, a new syndrome related to COVID 19 was first reported in a cohort of children from the UK and subsequently from many other countries all over the world. It is given many names – multisystem inflammatory syndrome in children (MIS-C) being the most widely accepted. Although initially it was thought to affect only children, now it has also been reported in adults, though much less commonly. Both World Health Organisation (WHO) and Centres for Disease Control and Prevention (CDC) has published diagnostic criteria for MIS-C (Table2).

The first publication of a neonate with SARS-CoV-2 presented on day seventeen of life with fever, cough, rhinorrhea and responded to supportive treatment. COVID-19 has also been reported in a 26 weaker preterm neonate. Neonates have reported fever, cough, rhinorrhea, apnoea, tachypnoea, tachycardia, vomiting, abdominal distention and diarrhea as presenting signs. They have been noted to have elevated CRP and deranged liver enzymes. Elevated myocardial enzymes have also been noted.

Our patient's mother was negative for COVID-19. None in the family had sign and symptoms suggestive of SARS-CoV-2. She was in two different hospitals previously and may have contacted the virus there. In the first two hospitals she was not tested for COVID.

She was diagnosed on day twenty-five of life. She had fever, tachypnoea, tachycardia with a fleeting maculo-papular erythematous rash. There is no prior documentation of dermatological manifestation in the neonatal population affected with SARS-CoV-2. Our index case initially had an erythematous rash with central clearing on day ten of life which may have been an early sign and was missed. She also received IVIG 1g/kg for thrombocytopenia. Whether this dose of IVIG inadvertently made a transient improvement of signs and symptoms due to COVID which subsequently worsened will remain a conjecture. She again developed erythematous maculo papular rash with necrotic changes on her 3rd week of life along with recurrence of fever. As the parents did not consent, biopsy could not be done.

She developed features suggestive of fulminant COVID-19 myocarditis characterised by poor cardiac contractility and elevated cardiac enzymes and NT-ProBNP and needed ionotropes twice. Mild myocarditis in neonates has been demonstrated in literature [9]. She also had markedly elevated inflammatory markers and D-dimer, thereby fulfilling all the criteria for MIS-C (Table 2). Possibly this is the first reported case of MIS-C in a newborn who had refractory myocarditis, dermatological involvement in the form of rash, gastrointestinal involvement in the form of diarrhoea, renal involvement in the form of high creatinine and may be CNS involvement in the form of convulsion. She needed prolonged steroid therapy and IVIG. Dermatological manifestation as a presentation of neonatal COVID-19 has also not been previously reported.

TABLE 2 Diagnosis of MIS-C

CDC Case Definition	WHO Case Definition
All 4 criteria must be met:	All 6 criteria must be met
1. Age <21 y	1. Age 0–19 y
2. Clinical presentation consistent with MIS-C, including all of the following:	2. Fever for ≥3 d
Fever: documented fever >38.0 C (100.4 F) or subjective fever for ≥24 h and ≥2 organ systems involved:	—
Cardiovascular (eg, shock, elevated troponin, elevated BNP, abnormal echocardiogram, and arrhythmia)	—
Respiratory (eg, pneumonia, ARDS, and pulmonary embolism)	—
Renal (eg, AKI and renal failure)	—
Neurologic (eg, seizure, stroke, and aseptic meningitis)	—
Hematologic (eg, coagulopathy)	—
Gastrointestinal (eg, abdominal pain, vomiting, diarrhea, elevated liver enzymes, ileus, and gastrointestinal bleeding)	—
Dermatologic (eg, erythroderma, mucositis, and other rash)	—
Laboratory evidence of inflammation, including but not limited to the following:	—
Elevated CRP, ESR, procalcitonin, and fibrinogen	—
Elevated D-dimer, ferritin, LDH, and IL-6	—
Neutrophilia and lymphocytopenia	—
Hypoalbuminemia	—
Illness requiring hospitalization	—
3. No alternative plausible diagnoses	3. Clinical signs of multisystem involvement (at least 2 of the following):
—	Rash; bilateral, nonpurulent conjunctivitis; or mucocutaneous inflammation signs (oral, hands, or feet)
—	Hypotension or shock
—	Cardiac dysfunction, pericarditis, valvulitis, or coronary abnormalities (including echocardiographic findings or elevated troponin and/or BNP)
—	Evidence of coagulopathy (prolonged PT or PTT; elevated D-dimer)
—	Acute gastrointestinal symptoms (diarrhea, vomiting, or abdominal pain)
4. Recent or current SARS-CoV-2 infection or exposure (any of the following):	4. Elevated markers of inflammation (eg, ESR, CRP, or procalcitonin)
Positive SARS-Cov-2 RT-PCR	—
Positive antigen test	—
Positive serology	—
Exposure to COVID-19 in the last 4 wk	—
—	5. No other obvious microbial cause of inflammation, including bacterial sepsis or toxic shock syndromes
—	6. Evidence of SARS-CoV-2 infection (any of the following):
—	Positive SARS-Cov-2 RT-PCR
—	Positive antigen test
—	Positive serology
—	Exposure to COVID-19 in the last 4 wk

AKI, acute kidney injury; ARDS, adult respiratory distress syndrome; BNP, B-type natriuretic peptide; CDC, Centers for Disease Control and Prevention; ESR, erythrocyte sedimentation rate; IL-6, interleukin 6; LDH, lactate dehydrogenase; PT, prothrombin time; PTT, partial prothrombin time.

Figure 1a : Erythematous rashes over legs on day 8

Figure 1b : Erythematous maculo papular rash in neck on day 24

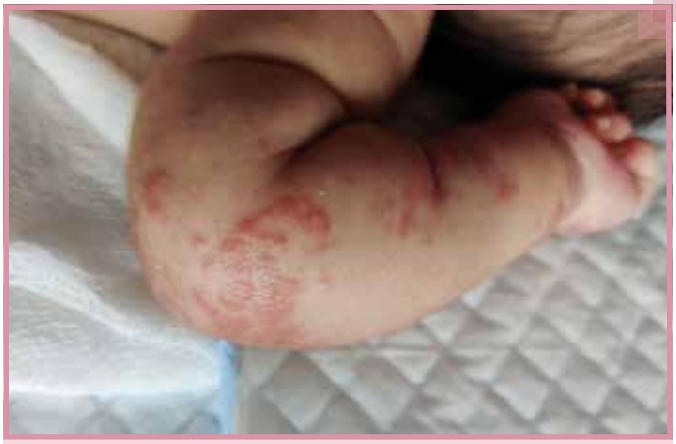

Figure 1c :Disseminated maculopapular rash over upper arm on day 24

Figure 1d : Necrotic patch over left groin on day 24

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Kikuchi Fujimoto Disease and Post - SARS COVID 19 Association

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Kikuchi–Fujimoto disease (KFD) was described in 1972 by Kikuchi and Fujimoto. KFD is a benign, self-limited lymphadenitis consisting of lymphadenopathy, fever, skin rashes, arthralgia, and hepatosplenomegaly, along with elevated levels of ESR, CRP, and LDH, lymphopenia, thrombocytopenia, and leucocytosis. The various aetiologies include viral infections and autoimmune diseases and recently, associations between COVID-19 and KFD were also reported. We discuss a case of KFD 6 week. after COVID-19 in a 10-yold girl who was admitted with high-grade fever for 3 weeks., bilateral, tender neck swelling, and macular rash off and on all over the body and face. There are very few case reports of associations between KFD and COVID-19 in the literature [1–3]. All the cases presented with fever and cervical lymphadenopathy within 2–3 month of COVID infection. Our patient had leukopenia, anaemia, and a raised ESR, similar to the cases described in the literature. All cases had raised LDH, including ours.

There is no specific diagnostic test for KFD. It is a diagnosis of exclusion. The diagnosis can be made only by histological examination of the involved lymph node and is characterized by a noncaseating necrotic area, karyorrhectic nuclear debris surrounded by mononuclear cells, particularly CD68+ histiocytes,

CD123+ plasmacytoid dendritic cells, and activated CD8+ T-lymphocytes. There is no specific treatment for KFD.

We report this case to highlight this sequalae of SARS COVID infection which will help in early diagnosis and the management of KFD.

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Publisher's Note Springer Nature Remains Neutral with Regard to Jurisdictional Claims in Published Maps and Institutional Affiliations.

Figure A : Paracortical zones of necrosis (H&E stain, X40)

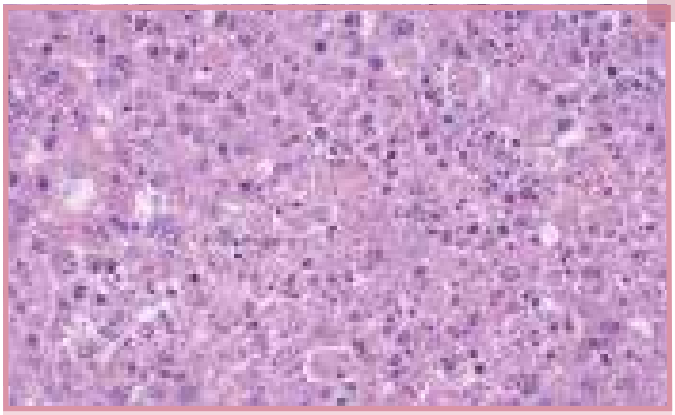


Figure B : Karyorrhectic debris with absence of neutrophils (H&E stain, X400)

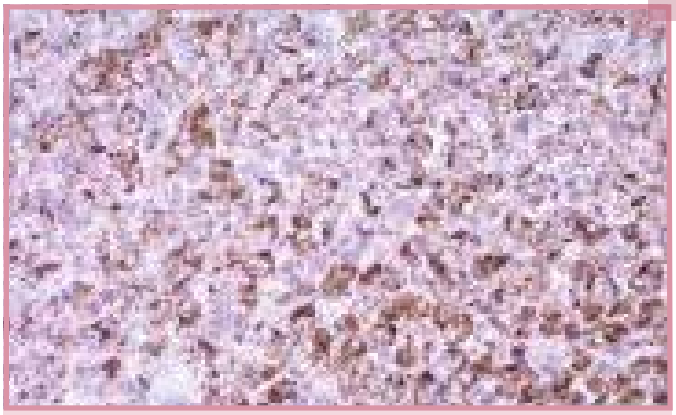
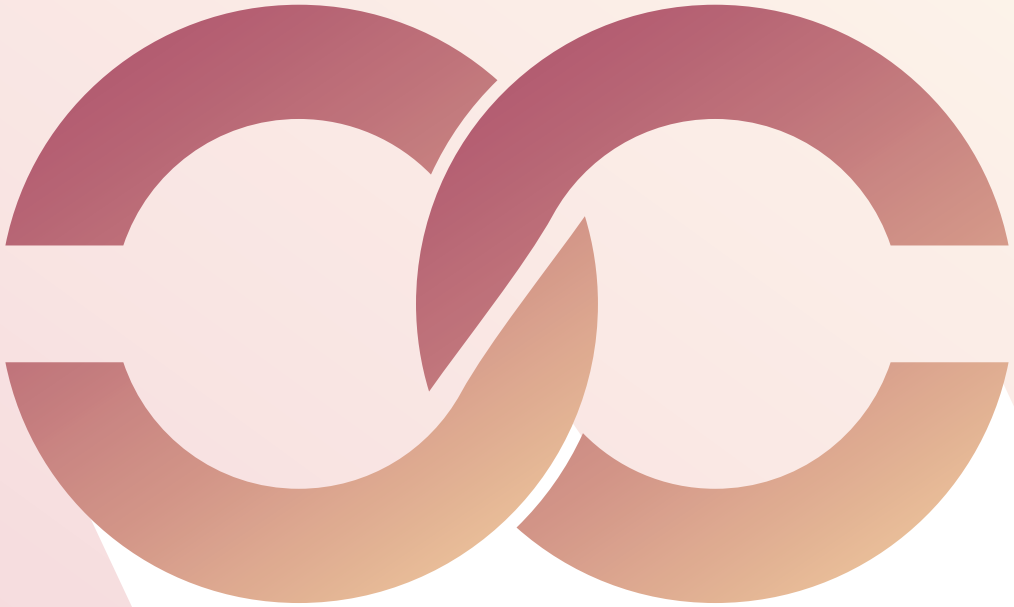


Figure C : Cd68 highlighting histiocytes (IHC, X400)

Figure D : Myeloperoxidase (MPO) stains histiocytes (IHC, X400)

Figure E : CD8 highlighting abundant T lymphocytes (IHC, X40)

Figure F : Cd4 weak positive in histiocytes and few T lymphocytes (IHC, X40)



RECOGNISING EXCELLENCE

Best Researcher Award

Winner

Dr Richie Gupta
Director and Head - Plastic,
Aesthetic and Reconstructive Surgery,
Fortis Hospital, Shalimar Bagh, New Delhi

Dr Rajat Gupta
Senior Consultant - Plastic,
Aesthetic and Reconstructive Surgery,
Fortis Hospital, Shalimar Bagh, New Delhi

Department of Plastic, Aesthetic and Reconstructive Surgery, Fortis Hospital, Shalimar Bagh Dr Richie Gupta and Dr Rajat Gupta are awarded 1st prize for Comparative Study of Urinary Complication Rates before and after the Incorporation of a Urethral Lengthening Technique during Masculinizing Genital Gender Affirmation Surgery.

Objectives

Masculinizing genital gender affirmation surgery (Mg GAS) consists of operative procedures designed to help the transition of transmen in their journey toward male gender role. Phalloplasty and urethral lengthening remain the most challenging of these surgeries, as the female urethra (4 cm long) must be lengthened to male dimensions (15–29 cm) with anastomosis at two sites, the native urethra/pars fixa urethra and the pars fixa urethra-penile urethra. As a result, there is a high incidence of urinary complications such as strictures and fistulae. Authors incorporated a urethral lengthening technique to reduce urinary complications in MgGAS. They compare the rates of urinary complications rates in cohorts before and after the introduction of this technique.

Materials and Methods

Authors have been performing phalloplasty since past 27 years, utilizing mainly free radial artery forearm flap (fRAFFp 431 cases) and pedicled anterolateral thigh flap (pALTp 120 cases). A retrospective review and comparison of urinary complications were performed before and after the introduction of their new technique since March 2017.

Results

There was a statistically significant reduction in the incidence of stricture with and without fistulae (25.94% with conventional and 4.17% with urethral lengthening technique p = 0.001) and fistulae alone (12.81% with conventional and 2.78% with urethral lengthening technique p = 0.011) in fRAFFp cases. In pALTp cases, the respective reductions were 43.08 to 17.07%, p = 0.006 (significant), and 13.85 to 4.88%, p = 0.197 (not statistically significant).

Conclusion

Over years, the rates of urinary complications in Mg GAS have remained constant, varying from 25 to 58% for strictures and 17 to 75% for fistulae as noted by many authors. Authors noted that in most of their cases, strictures occurred at distal pars fixa urethra (DPFU)-penile urethra anastomosis and incorporated a urethral lengthening technique, which lengthens the DPFU by 3 to 5 cm at this anastomotic site, thus significantly reducing the anastomotic tension and the rate of urinary complications.

Best Researcher Award

1st Runner - up

Dr Rajesh Gupta
Associate Director - Pulmonology & Critical Care, Fortis Hospital, Greater Noida

Dr Sunny Kumar
Consultant - Pulmonology and Critical Care, Fortis Hospital, Noida

Dr Saurabh Mehra
Consultant - Pulmonology and Critical Care, Fortis Hospital, Noida

Dr Onkar Jha
Associate Consultant - Pulmonology & Critical Care, Fortis Hospital, Greater Noida

Department of Pulmonology and Critical care, Fortis Hospital, Noida - Dr Rajesh Gupta and Dr Sunny Kumar, Dr Saurabh Mehra and Dr Onkar K Jha are awarded 2nd prize for their article Helmet NIV in Acute Hypoxemic Respiratory Failure due to COVID-19: Change in PaO2/FiO2 Ratio a Predictor of Success published in Indian Journal of Critical Care Medicine

In acute respiratory failure due to severe coronavirus disease 2019 (COVID-19) pneumonia, mechanical ventilation remains challenging and may result in high mortality. The use of non-invasive ventilation (NIV) may delay required invasive ventilation. increase adverse outcomes., and have a potential aerosol risk to caregivers.

Data of 30 patients were collected from patient files and analyzed. Twenty-one (70"o) patients were weaned successfully after helmet-NIV support (NIV success group). and invasive mechanical ventilation was required in 9 (30%) patients (NIV failure group) of which 8 (26.7%) patients died. In NIV success vs failure patients. the mean baseline PaO/Fi02 ratio (PFR) (147.2 + 57.9 vs 156.8 + 59.0 mm Hg: p- 0.683) and PER before initiation of helmet (132.3 + 46.9 vs 121.6 + 32.7 mm Hg: p = 0.541) were comparable. The NIV success group demonstrated a progressive improvement in PFR in comparison with the failure group at 2 hours (158.8 + 56.I vs 118.7 t 40.7 mm Hg: p = 0.063) and 24 hours (PFR-24) (204.4 + 94.3 vs 121.3 + 32.6: p = 0.016). Helmet interface NIV may be a safe and effective tool for the management of patients with severe COVID-19 pneumonia with acute respiratory failure. More studies are needed to further evaluate the role of helmet NIV especially in patients with initial PFR <150 mm Hg to define PFR/dPFR cutoff at the earliest time point for prediction of helmet-NIV success.

Best Researcher Award

Runner - up

Dr Murali Manohar
Senior Consultant -
Cardiothoracic Vascular Surgery,
Fortis Hospital, Banerghatta Road,
Bengaluru

Dr Siddhu S Neginahal
Senior Perfusionist -
Cardiothoracic Vascular Surgery,
Fortis Hospital, Banerghatta Road,
Bengaluru

Dr Sudarshan GT
Consultant -
Cardiothoracic Vascular Surgery,
Fortis Hospital, Banerghatta Road,
Bengaluru

Department of Cardiothoracic and Vascular Surgery - Dr Murali Manohar, Siddhu S Neginahaland and Dr Sudarshan GT - Fortis Bannerghatta Road, Bangalore are awarded 3rd prize - Hemadsorption in complex Cardiac Surgery - single-center retrospective observational study of patients undergoing complex cardiac surgery published in Journal of Clinical medicine

Cardiac surgery may evoke a generalized inflammatory response, typically magnified in complex, combined, redo, and emergency procedures with long aortic cross-clamp times. Various treatment options have been introduced to help regain control over post-cardiac surgery hyper-inflammation, including hem adsorptive immunomodulation with CytoSorb®. (2) Methods: We conducted a single-center retrospective observational study of patients undergoing complex cardiac surgery. Patients intra-operatively treated with CytoSorb® were compared to a control group. The primary outcome was the change in the vasoactive-inotropic score (VIS) from pre-operatively to post-operatively. (3) Results: A total of 52 patients were included in the analysis, where 23 were treated with CytoSorb® (CS) and 29 without (controls). The mean VIS increase from pre-operative to post-operative values was significantly lower in the CS group compared to the control group (3.5 vs. 5.5, respectively, $p = 0.05$). In-hospital mortality in the control group was 20.7% (6 patients) and 9.1% (2 patients) in the CS group ($p = 0.26$). Lactate level changes were comparable, and the median intensive care unit and hospital lengths of stay were similar between groups. (4) Conclusions: Despite notable imbalances between the groups, the signals revealed point toward better hemodynamic stability with CytoSorb® hemadsorption in complex cardiac surgery and a trend of lower mortality.

Best Researcher Award

Runner - up

Dr Ishita B Sen
Senior Director and Head - Nuclear Medicine,
Fortis Memorial Research Institute, Gurugram

Dr Parul Thakral
Assistant Manager - Clinical Research, Nuclear Medicine,
Fortis Memorial Research Institute, Gurugram

Dr Subha Shankar Das
Consultant - Nuclear Medicine,
Fortis Memorial Research Institute, Gurugram

Department of Nuclear Medicine- Fortis Memorial Research Institute, Gurgaon - Dr Subha Shankar Das, Dr Parul Thakral, and Dr Ishita B Sen are awarded 3rd prize –- Feasibility of 18 F-FDG Labelling of Leucocytes in a centre without onsite Cyclotron and monitoring of Radiation Dose of Occupational Worker in the Labelling Procedure published in Journal of Cancer Biotherapy and Radiopharmaceuticals

Differentiation of infection from sterile inflammation is still a major concern for clinicians. The18F-WBC positron emission tomography/computed tomography scan has been considered a promising tool for accurate diagnosis of infection owing to its high specificity, but it renders the availability of a medical cyclotron a necessity. The aim of the present study was to determine the feasibility of labelling leukocytes and establish the protocol in a centre without the availability of an on-site medical cyclotron. The secondary aim was to monitor radiation doses to occupational workers involved in labelling of leukocytes with18F-FDG.

Conclusions

Labelling of leukocytes with18F-FDG is possible at a tertiary nuclear medicine setup without the availability of an on-site medical cyclotron, with reasonable labelling efficiency of 78.01% ± 6.99%. In addition, in-house labelling of leukocytes with18F-FDG is safe and the radiation doses incurred by the personnel during the labelling procedure are well within the occupational dose limits established by the national regulatory authority.

Golden Hands Award

Dr Nishit Sawal
Consultant - Neurology
Fortis Hospital, Mohali

Department of Neurology- Dr Nishit Sawal, Fortis Hospital, Mohali is awarded 1st prize for his case DBS (Deep Brain Stimulation) of Bilateral Ventro-Medial Hypothalamus for Pathological Hypersexuality – **World's First Case**

Miss X, 27-year-old female; At the age of 5 years, patient suffered a febrile encephalopathy. Later at the age of 13 after attaining menarche, she developed excessive and abnormal sexual urge. Medical treatment, multiple ECT's, sex hormone therapy – all proved to be of no benefit at all. DBS of both Ventro-medial Hypothalami was done in May'2022. Boston Scientific Vercise leads were used. Switching both right/left VMH led to complete control of her sexual symptoms. Switching on right NA also led to marked reduction in her irritability while sexual urge remained totally negated. This is the first case of VMH DBS in the world. The girl had a complete remission of her symptoms to the point where all her psychiatric medicines were also completely stopped. This case illustrates the efficacy of DBS for such disorders.

- Also, it has important medico-legal ramifications as to whether all serial sexual offenders/rapists should be offered VMH DBS and post VMH DBS, can they be given paroles or prison breaks? More research is needed to clarify these points. Western psychiatrists/lawyers have been arguing for some medical procedures to be conducted upon serial sexual offenders with impulse control disorders so that their incarceration can be made more humane. This case - the first in the world – provides a blueprint for the same

Golden Hands Award

Dr Satish Javali
Consultant - Cardiothoracic Vascular Surgery
Fortis Hospital, Mulund

Dept of Cardiovascular Surgery - Fortis Hospital, Mulund, Mumbai Dr Satish Javali is awarded 2nd Prize for his article Eroding Pseudo Aneurysm of Ascending Aorta - Published in Fortis Clinical connect issue 6 Sept 22 page55

A case of large mass (pseudoaneurysm) arising from the ascending aorta 2 cm above the aortic valve and extending into the right hemithorax. Anticipating difficulty in performing a redo sternotomy, a 8mm graft was sutured to the right axillary artery and cannulated with a 22 Fr arterial cannula, the right femoral vein was cannulated with a 25 Fr venous ECMO cannula and bypass was commenced. Its a rare and delayed presentation of aortic Pseudoaneurysm.

The intra-op decisions to tackle aortic rupture and unique way to achieve cardiac arrest are not literature guided but innovated on table. Such a large Pseudoaneurysm repair is associated with considerable morbidity and mortality. In this case patient recovered without any neurological complications, bleeding and other post cardiac surgical complications. With meticulous planning, involvement of the whole team- the anaesthesiologists, the perfusionists nurses and assistants along with smooth execution of circulatory arrest and sheer luck, we could achieve a good result.

Dr Keshava R
Senior Director - Cardiology,
Fortis Hospital, Cunningham Road, Bengaluru

Dr Anusha Rao
Associate Consultant - Cardiology,
Fortis Hospital, Cunningham Road, Bengaluru

Dept of Cardiology - Dr Keshava R and Dr Anusha Rao - Fortis CG Road-*Inadvertent Extraction of a Fully Deployed Stent during Retrieval of an Undeployed Stent by Indigenous Handmade Snare*

62-year-old doctor was referred from a peripheral centre. He had undergone emergency angioplasty to LAD two months back. He was taken up for a staged PTCA of circumflex. Patient was taken electively for PTCA of circumflex artery, radial procedure was started. We brought this patient after 48hrs because the dye load was more, we did the OCT. We found the left main and LAD Stent was fully expanded. We struggled to go circumflex. So we didn't do the OCT of circumflex artery. However, we couldn't find the thin wire hanging on the OCT. But it's clearly evident on the 2D ECHO and the patient is doing extremely well. What is unique about this case is because of non-availability of required snare, you can do manufacture your own snare on table with our own indigenous innovative technique. It's already been described by somebody. We could do it at night 8PM to 10 PM, make our own snare and snare the stent and prevent the obvious catastrophy should have happened if this stent was not retrieved.

Golden Hands Award

Dr Gopi
Senior Director - Cardiology,
Fortis Hospital, Cunningham Road, Bengaluru

Dr Harsha
Associate Consultant - Cardiology,
Fortis Hospital, Cunningham Road, Bengaluru

Dept of Cardiology – Fortis Hospital Cunningham Road - Dr Gopi and Dr Harsha are awarded 3rd Prize- A Teamwork that Made the Difference

Mr Mohammed a 33-year-old well-built healthy male had undergone an arthroscopic surgery to the right knee 10 days ago and was immobile for the last few days. Patient presented to the ER with severe breathing difficulty and chest pain of an hour duration. On arrival at ER patient's respiratory rate was 60 per minute, heart rate was 141 beats per minute and SP 02 was 86%. Patient was started on oxygen. Patient's ECG showed sinus tachycardia with RBBB. All of a sudden patient developed bradycardia and impending cardiac arrest, immediately ACLS protocol was started and patient was intubated. ROSC achieved in 15 minutes again patient had three further episodes of cardiac arrest but could be resuscitated immediately. In the meanwhile, an emergency echocardiography was done which showed a dilated right atrium and right ventricle with the PA pressure of 60mmhg. A clinical diagnosis of massive pulmonary embolism with cardiogenic shock was made. Patient remained hypotensive in spite of inotropes. As patient unstable he could not be shifted for a CT pulmonary angiogram to confirm the diagnosis. The same was discussed with the relatives and a decision was made to empirically thrombolyse the patient with the Tenecteplase 50 mg. Patient was thrombolysed and shifted to the CCU. Patient gradually became stable hemodynamically. Next day biochemistry revealed patient has acute kidney injury and shock liver with grossly increased liver enzymes. Patient was gradually weaned of the ventilator and extubated the next day. Patients AKI and liver dysfunction gradually improved over the next week. Repeat echocardiography showed reduction in size of RA and RV and PA systolic pressure reduced to 43 mmHG. Patient was eventually discharged after eight days and presently on follow-up patient is back to normal and repeat echocardiography showed that RA and RV is normal in size with normal pulmonary artery pressures. Prompt effective resuscitative measure by the ER and code blue team was critical in saving the patient's life.

Fortis Mulund Felicitates Rotary Club of Mumbai-Mulund South for Helping Fund Over 150 Paediatric Cardiac Surgeries

On April 8, 2023, Fortis Hospital, Mulund, honoured members of the Rotary Club of Mumbai-Mulund South for their contribution and financial support towards Paediatric Cardiac surgeries for underprivileged children. The Rotary team was felicitated by Dr S. Narayani, Business Head-Fortis Hospitals, Maharashtra. The Paediatric Cardiac team at Fortis Hospital, Mulund, has performed 151 surgeries in children in the age group of 6 months to 17 years, with the support of the Rotary Club of Mumbai-Mulund South.

Dr Narayani said, "Our heartfelt gratitude to Rotary members helping us drive these lifesaving surgeries for children from disempowered backgrounds. We at Fortis believe that every child deserves access to quality healthcare, regardless of their socioeconomic background, and this is a positive step on that front." The event was attended by Rotary members, paediatric cardiac patients with their parents, and the Paediatric Cardiac team, including Dr Dhananjay Malankar, Consultant-Paediatric Cardiac Surgeon, Dr Swati Garekar, Consultant-Paediatric Cardiologist, Dr Sachin Patil, Lead Paediatric Anaesthetist & Intensivist, Dr Shivaji Mali, Paediatric Anaesthetist & Intensivist, Dr Shyam Dhake, Consultant-Paediatric Anaesthesiologist & Intensivist, Dr Bharat Soni, Consultant Paediatric Cardiac Surgeon, and Ms Manasi, Chief Paediatric Cardiac ICU Nurse & her team, at Fortis Hospital Mulund.

(R-L) Ms Rinku Mavani, Business Partner-Sales, Fortis Hospitals Mumbai with Rtn Praveen Khattar

(L-R) Dr S. Narayani, Business Head-Fortis Hospitals Maharashtra with Rtn Shailesh Parekh

(Seated C) Dr S. Narayani with the paediatric cardiac team, HODs, clinicians, patients & their families along with Rotary Club representatives

Ms Gowri S. Jagadesan of Fortis BG Road, Bengaluru, Participates as National Trainer for 'Breastfeeding and Infant & Young Child Feeding Counsellor Course'

Ms Gowri Sayee Jagadesan, Nurse Educator, Fortis Hospital, Bannerghatta Road, Bengaluru, successfully participated as a National Trainer for the Breastfeeding and Infant & Young Child Feeding Counsellor Course, the 4-in-1 training organised by a prestigious hospital in Hyderabad along with Breastfeeding Promotion Network of India (BPNI) during March 20-26, 2023. She participated as a BPNI National Trainer IYCF along with few other notable IYCF National Trainers. A total of 26 participants, including paediatricians, physiotherapists, nurses, nutritionists, lactation counsellors, pharmacists and wellness consultants underwent the course.

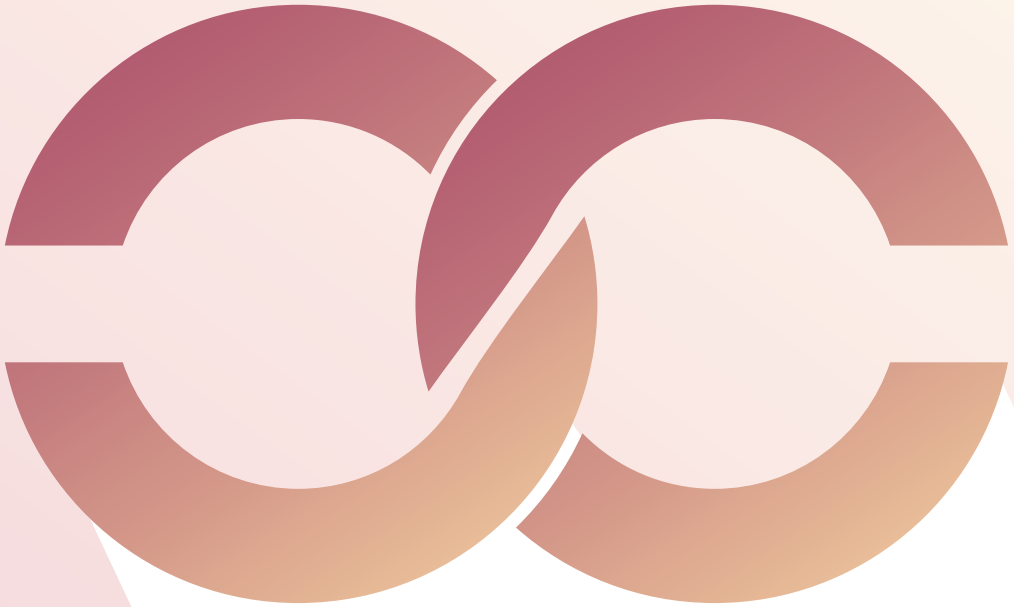






BPNI's Certificate Course on
"Breastfeeding & Infant and Young Child Feeding Counsellors"
Date: 20-26 March 2023


NILOUFER HOSPITAL: INSTITUTE OF CHILD HEALTH, GOVERNMENT OF TELANGANA
&
BREASTFEEDING PROMOTION NETWORK OF INDIA (BPNI)
Certificate Of National Trainer
Gowri Sayee Jagadesan
"Breastfeeding & Infant and Young Child Feeding Counsellor"



ADVANCES IN DIAGNOSTICS

Ultrasound Examination of the Lung

Dr Krishan Chugh
Principal Director and Head - Paediatrics and PICU, Fortis Memorial Research Institute, Gurugram

Dr Neetu Talwar
Additional Director - Paediatrics Pulmonology, Fortis Memorial Research Institute, Gurugram

Keywords: USG lung, PLUS, Community Acquired Pneumonia (CAP)

Point of care ultrasound examination is gaining popularity in recent times. It essentially means that the ultrasound machine would be used by the clinician himself and not the radiologist at the point where the patient needs care. It is used both for diagnosis as well as for helping in procedures.

However, lung ultrasound is a somewhat different story. It was always believed that the ultrasonic waves do not pass through the ribs and in the intercostal space they would encounter air of the lungs which is also a poor conductor of ultrasonic waves. So, when an ultrasound probe is kept on the chest, the waves would not pass through the ribs and when they pass through intercostal space we would see only some artefacts unless fluid is collected in the lung/pleura (pleural effusion). It was Lichtenstein who looked at these artefacts carefully, indeed very carefully and concluded that artefacts also have a pattern that varies with the condition of the lung inside [(1).] Studies of lung ultrasound have shown that the patterns are quite different in patients with pneumonia, atelectasis, pneumothorax, pulmonary edema, ARDS and so on [(2).]

In recent years, ultrasound of the lung has been compared with x-rays in the diagnosis of community acquired pneumonia. Most of the studies were done in adults. At Fortis Memorial Research Institute, Gurgaon, we conducted study in small children who clinically satisfied the criteria laid down by WHO for the diagnosis of pneumonia (N = 148) [(3).] These children were subjected to both x-ray as well as ultrasound of lung done by the clinician. The clinician performing ultrasound examination was not aware of the x-ray findings, even if x-ray had been done before ultrasound. A systematic approach was followed for complete evaluation of the lungs on both sides, anteriorly as well as posteriorly [(Figure1).]

On comparison, it was concluded that paediatric lung ultrasound (PLUS) is equally good or actually somewhat better in detecting community acquired pneumonia. In the cases where there was a discrepancy between the diagnosis made by ultrasound and x-ray, CT scan was performed taking that as gold standard. Overall, it was seen that in patients where there was a discrepancy, CT scan more often agreed with ultrasound findings than with x-ray [(Figure2).] This study was done in patients who were hospitalised.

Radiation exposure is an important consideration in children with regard to the damage that it can do to their reproductive health as well as their higher chances of developing malignancy in the long life that lies ahead [(4).] Ultrasound, being a non-radiating modality, becomes an obvious choice for diagnosis of community acquired pneumonia in children over x-ray. However, it must be conceded that there are some areas of the lung which are not easily evaluated by ultrasound examination for example deep medial parts of the lung. It has been shown in several studies in the past that the learning curve for the sound is short and simple. A short course under the care of a trained clinician or a radiologist is sufficient. Almost ubiquitous availability of ultrasound in all clinical areas of the hospitals is an additional benefit. Most of the hospitals have a limited number of mobile x-ray machines and it may not be always safe, or feasible for a patient to be transported to the x-ray department.

Hence, the scorecard is clearly in favor of ultrasound for detection of community acquired pneumonia and probably also in following up its course over the next few days [(5).]

Figure 1. (a, b) : Areas of thoracic region to be scanned by ultrasound: 1 and 2, anterior superior, anterior inferior; 3 and 4, lateral superior and lateral inferior; 5 and 6, posterior superior and posterior inferior; AAL, anterior axillary line; PAL, posterior axillary line.

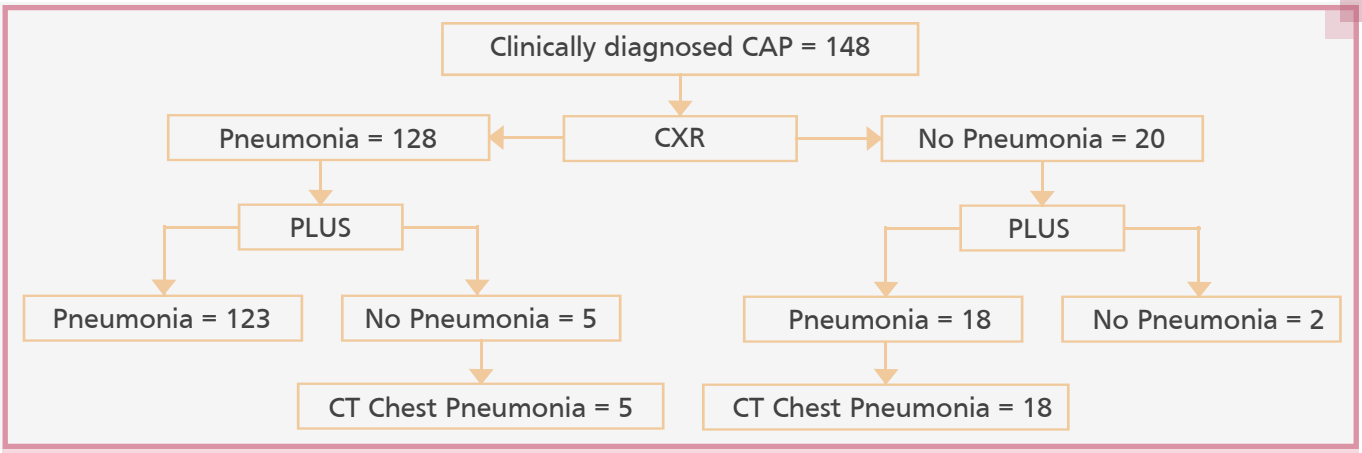


Figure 2 : Details of Discordant Patients. CXR – chest r ray; PLUS – paediatric lung ultrasound; CAP – community acquired pneumonia.

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Computed Tomographic Evaluation of Congenital Left Ventricular Outflow Obstruction

Source: https://dx.doi.org/10.2174/1573403X19666230525144602

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Congenital left ventricular outflow obstruction represents a multilevel obstruction with several morphological forms. It can involve the sub valvular, valvar, or supra valvular portion of the aortic valve complex, and may coexist. Computed tomography (CT) plays an important supplementary role in the evaluation of patients with congenital LVOT obstruction. Unlike transthoracic echocardiography and cardiovascular magnetic resonance (CMR) imaging, it is not bounded by a small acoustic window, needs for anaesthesia or sedation, and metallic devices. Current generations of CT scanners with excellent spatial and temporal resolution, high pitch scanning, wide detector system, dose reduction algorithms, and advanced 3-dimensional postprocessing techniques provide a high-quality alternative to CMR or diagnostic cardiac catheterization. Radiologists performing CT in young children should be familiar with the advantages and disadvantages of CT and with the typical morphological imaging features of congenital left ventricular outflow obstruction.

Keywords: left ventricular outflow obstruction, congenital heart disease, computed tomography, coartation of aorta, aortic stenosis, transthoracic echocardiography

Recent Advances in Genomic Testing for Paediatrics

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Background

Incidence of congenital malformations in general population is 3-4%. Incidence of chromosomal abnormalities at birth is 0.6%. Incidence of single gene disorders at birth is 1-2%. Diagnosis and management of such disorders is difficult and challenging. There has been a paradigm shift in genetic testing in last decades. A clinician of 21st century needs to understand the basics of medical genetics and technical advances so as to improve the standard of patient care.

Common Indications of Genetic Testing for Paediatricians

1. A child with congenital malformations
2. Dysmorphic child
3. A child with intellectual disability
4. A child with autism spectrum disorder
5. A newborn with ambiguous genitalia
6. A child with Short stature
7. A child with motor delay
8. A child with metabolic disease
9. A child with seizures
10. A child with hematological diseases e.g. Thalassemia, Hemophilia
11. A child with Cancer
12. A child with vision problems
13. A child with deafness
14. A child with ataxia

Genetic tests look at one or a few genes at a time. Genomic tests can look at hundreds or thousands of genes at a time, yielding much more information in one single test.

Genetic testing basically involves detection of chromosomal abnormalities, CNV (Copy number variants) single gene disorders and epigenetic.

Chromosomal disorders include

- Numerical (aneuploidy)
- or Structural.

Numerical abnormalities include loss or gain of a large part of one or more chromosomes e.g. Down syndrome, Trisomy 18, Trisomy 13.

Structural abnormalities include balanced translocations, inversions and insertions.

Loss or gain of smaller regions of a chromosome, known as copy number variations (CNV), usually involve more than one gene and are implicated in many human diseases Single gene disorders are caused by sequence variation in the genes.

The testing for genetic disorders is basically checking for chromosomal abnormalities. CNV's and single gene disorders. Various techniques for testing these are based upon whether cytogenetic or molecular tests are indicated.

Karyotyping is the gold standard for detecting chromosomal aneuploidies.

Chromosomal microarray analysis (CMA) is now widely used to detect chromosomal abnormalities and CNV's.

FISH is only indicated if the diagnosis is known already e.g. Prader Willi syndrome, DiGeorge syndrome.

Cytogenetic Testing	Molecular Testing
Karyotype	PCR, RFLP, ARMS....
Fluorescent in situ hybridization	Sanger sequencing
Chromosomal microarray	Next Generation Sequencing

Salient Features of Cytogenetic Testing

Karyotype	FISH- Fluorescent in Situ Hybridization	Chromosomal Microarray
Resolution: 5-10 Mb Needs expert eyes for interpretation. Time consuming (2-3 weeks for results). Cannot detect LOH, UPD.	Resolution: average 80kb -1 Mb for constitutional aberrations. Locus specific- we need to know the targeted region. FISH is considered a validation NOT a screening test. Fluorescence microscope required.	Resolution: down to 50kb (or less) 100% sensitivity and specificity for 400kb CNVs. Results in 3 days. User friendly software for data interpretation. SNP probes allow detection of LOH, UPD, parent of origin and consanguinity.

G-banded karyotyping is the most common approach for the detection of genomic alterations.

However, Karyotype is unable to detect genomic changes of less than 5 Mb. The ability of FISH to detect cryptic chromosomal rearrangements exceeds the resolution of any form of cytogenetic banding techniques. However, conventional FISH does not allow a comprehensive evaluation of the whole genome. Thus, FISH provides a high-resolution analysis of only targeted locations.

Chromosomal Microarray

CMA, otherwise known as genomic microarray, enables the study of chromosomes at a higher resolution as compared to traditional karyotyping. It has replaced karyotyping as the first-tier investigation of children with intellectual disability, multiple malformations and autism as per ACMG criterion [2,3]. CMA offers a much higher diagnostic yield (15%–20%) for individuals with unexplained DD/ID, ASD, or MCA.

CMA- Basic Concepts

DNA microarray is like a computer chip and contains a large assembly of DNA fragments spotted onto a solid surface such as glass. These DNA fragments are DNA probe and size of DNA probe is - 25 to 80 base pairs. Human genome contains over 3 billion base pairs, so a DNA microarray would require multimillion probe in order to comprehensively cover the entire genome. As such, arrays used for clinical purposes do not necessarily need to have representation of every human sequence and are intelligently designed to cover coding or noncoding functional genes. They are spread across the genome at regular intervals (form the 'backbone' and defines the resolution of CMA) and are usually enriched for regions of clinical interest. They are designed to detect CNVs or single nucleotide polymorphisms (SNPs) or both.

Types of CMA

There are two CMA techniques used for identifying chromosomal imbalance:

- Comparative genomic hybridization (CGH) and
- Single nucleotide Polymorphism (SNP)

Microarray-based Comparative Genomic Hybridization (aCGH)- Oligoarray detects genome-wide microscopic and sub-microscopic copy number variants (CNVs) less than 100 kb. Single nucleotide polymorphism (SNP) arrays

SNPs provide more even genome coverage and improved detection of CNVs compared to aCGH, triploidy, UPD, Mosaicism Most commercially available platforms are hybrid arrays and contain oligonucleotide probes for detecting both CNVs and SNPs.

Interpretation

The variants identified are critically evaluated based on their size, gene content and published reports in literature. The databases used for CNV interpretation are Decipher, UCSC genome browser, Clinvar etc.

The CNVs are classified (based on American College of Medical Genetics and Genomics (ACMG) criteria)

1. Pathogenic,
2. Benign or
3. Variant of uncertain significance (VOUS)

VOUS are variants, which are not directly linked to the patient's phenotype but have some evidence for causation. Testing of parents may be required to ascertain the significance of the variant.

Before ordering CMA, one should know the design and resolution of the testing platform and the genomic regions covered. Most of the commercial platforms available have probes for known microdeletion/ duplication syndromes along with genome wide probes for other clinically significant

CNVs. Microarray can be low resolution or high resolution depending upon the number of probes used. A typical 750 K microarray is sufficient for routine testing of a child with intellectual disability, congenital malformation and autism.

Both pre-test counselling for the diagnostic sensitivity (detection rate 15-20% higher compared to routine Karyotype) and limitations) and post-test counselling are essential.

Limitations of CMA

- Cannot detect monogenic disorders
- Cannot detect balanced translocations
- Cannot detect mosaicism <10-15%
- Variant of uncertain significance (VOUS)
- Need of Parental studies

Molecular Tests for Monogenic Disorders Include

1. Sanger sequencing
2. ARMS- PCR, RFLP etc.
3. MLPA
4. Next gene- ration sequencing (NGS)

Next - Generation Sequencing

NGS, also known as massively parallel sequencing or deep sequencing, is a high throughput sequencing technology which allows simultaneous sequencing of millions of DNA base pairs at a comparatively lower cost and higher speed. Exomes comprise only 1% of 6.2 billion base pairs in human DNA, which code for proteins.

NGS based analysis includes three major groups: Clinical exome sequencing (CES); Whole exome sequencing (WES); Whole-genome sequencing (WGS).

1. Clinical exome
2. Whole exome
3. Whole genome
4. Gene panel

Exons are the protein-coding region of the genome, which make up 1% to 2% of the total genome, but more than 85% of all disease-causing mutations are reported in these regions. Exome sequencing cover only exons. Genome sequencing covers both introns and exons.

Whole exome sequencing is mainly being used for suspected monogenic disorders. A trio analysis (child and parents) is the most ideal approach to genetic testing.

Clinical Exome Sequencing

In clinical exome sequencing, only those genes are included which are known to be disease causative and the total number of genes in this would vary depending upon the laboratory, and generally, it can cover 5000- 7000 genes.

Whole Exome Sequencing

In whole exome sequencing, all exons are tested and testing would include around 20000 genes.

Whole Genome Sequencing

Whole genome sequencing involves testing of whole exons and introns and currently mainly used as a research tool.

Interpretation of NGS Based Tests

The variants are filtered to narrow down to a single variant that is most likely to explain the disease or phenotype. Several computational tools are now available to predict the effect of a change in the nucleotide sequence of a gene e.g. Mutation taster, the sorting (also popularly called filtering) is also aided by published databases of normal variants and disease-causing variants. In 2015, ACMG published guidelines for interpretation of sequence variants and categorized them into five categories, i.e., pathogenic, likely pathogenic, benign, likely benign and VOUS. The results are then correlated with clinical features and communicated to the patient.

A patient should be referred to a trained clinical geneticist in case of abnormal results. The clinical geneticists are trained to correlate the NGS results clinically and do three generation pedigree analysis. They may need to do further biochemical testing or other investigations e.g. Xray , MRI to confirm the pathogenicity of the NGS reports. A correct diagnosis helps in etiological diagnosis and genetic counselling for recurrence risk.

CMA is ordered first in a case with intellectual disability or congenital malformations/ autism. NGS should be the first choice if there is a family history or consanguinity or suspected single gene disorders.

Conclusions

- Genomic testing is rapidly becoming an integral part of clinical practice
- Chromosomal microarray, exome sequencing and whole genome sequencing using NGS techniques are powerful methods to investigate variations in human genome

- Hence, its important to be well versed with these tests so as to improve the evidence based health care standards

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Abernethy Malformation: A Comprehensive Review

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Abstract

Abernethy malformation is a rare condition in which portomesenteric blood bypasses the liver and drains into a systemic vein through a partial or complete shunt. It is categorized into 2 types on the basis of the shunt pattern between the portal vein and a systemic vein. Abernethy malformation is associated with multiple congenital anomalies and acquired complications. A detailed understanding of the anatomy and embryology is a prerequisite to interpret imaging findings. Computed tomography and magnetic resonance angiography can delineate the shunt anatomy and evaluate concomitant malformations. It is essential to differentiate Abernethy malformation from intrahepatic portosystemic shunts and acquired extrahepatic portosystemic shunts. Mild metabolic abnormalities are treated with dietary modifications and medical therapy. Definitive treatment is done in symptomatic patients. Generally, type I Abernethy patients undergo shunt occlusion by surgery or transcatheter coiling.

Main Points

- Type I Abernethy is characterized by atretic intrahepatic portal venous branches and complete extrahepatic shunting of portal blood into a systemic vein, while type II is characterized by hypoplastic intrahepatic portal venous branches with partial extrahepatic shunting of portal blood into a systemic vein.
- Abernethy malformation is associated with multiple congenital malformations and acquired complications.
- It is essential to differentiate Abernethy malformation from congenital intrahepatic and acquired extrahepatic shunts.
- Multipurpose catheters should be considered when typical catheters fail to identify the RAV or when cranially oriented RAVs are identified but cannot be sampled with typical catheters.

Conclusion

Abernethy malformation is a rare anomaly with multiple clinical associations. Most often, children present with dyspnea, encephalopathy, and abdominal complaints. The purpose of imaging is to identify and classify the shunt, with identification of accompanying anomalies. It is essential to distinguish this entity from intrahepatic shunts and acquired extrahepatic shunts. Careful monitoring is recommended if the patients are asymptomatic or have mild metabolic abnormalities. Any complication warrants appropriate therapeutic intervention. Radiologists must be familiar with imaging features of this rare anomaly for early diagnosis and therapeutic guidance, leading to better patient outcome.

Bronchoscopy in PICU

Source : doi: 10.4103/IJPCC.IJPCC_35_23

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Keywords: Flexible bronchoscopy, PICU, BAL

Patients in the paediatric or neonatal intensive care units usually are critical, very often with some degree of respiratory compromise requiring mechanical ventilation – invasive or non-invasive. The procedure of flexible bronchoscopy (FB) poses a challenge in these children because the airway is going to be obstructed for that short duration even if the smallest of bronchoscopes is used, which may not be easily tolerated by the patient whose oxygenation, ventilation, circulation or CNS is compromised.

It is important to understand the pathophysiological changes that insertion of a bronchoscope would result in, so that adequate preventive measures can be taken accordingly. The most important consequence would be an increase in the airway resistance requiring adjustment of the ventilator flow in an intubated child. Instillation of fluids into the lungs for getting a bronchoalveolar lavage (BAL) would result in some compromise of the compliance of the lung. These two changes would require adjustment of the ventilatory settings. Further, changes in the pressure in the lungs and airways may result in increased intracranial pressure as well as some hemodynamic changes.

Access to the lower airways can be obtained via the endotracheal tube, tracheostomy tube, Laryngeal mask airway (LMA) or directly through the mouth or the nose in an non-intubated child. During the procedure, the child should be adequately sedated, and in many situations even temporarily paralyzed. The bronchoscopist should have a plan ready beforehand so that the time required is minimized and yet the maximum information required is collected and if any procedure is pre planned that also is completed.

Indications of Bronchoscopy in PICU / NICU

Diagnostic Indications

FB is used as the procedure of choice in intensive care units for various reasons. The most common reason for performing FB in PICU is for obtaining broncho-alveolar lavage (BAL) sample. A BAL obtained from the suspected pathological area yields a lot of information in children with lung infections. The BAL obtained this way can be subjected to the standard microscopic Gram stain and cultures[(2),] In recent times. Multiplex PCR has been added to the armamentarium of the paediatric intensivist that can read the DNA imprints of many viruses and bacteria. Further, it can also give a list of the genes that may be present which result in antimicrobial resistance. The clinician can target his treatment according to these results. Non-infective pulmonary causes like pulmonary hemorrhage, aspiration pneumonia, lipoid pneumonia may give characteristic features on BAL sample.

X-ray findings of atelectasis or hyperinflation can be assessed using FB. Airway assessment stays the other important indication in cases of persistent noisy breathing caused by stridor or wheeze. Conditions like laryngomalacia, subglottic stenosis [(Figure1),] vocal cord paralysis, hemangioma, laryngeal anomalies (cyst, web, cleft) can cause persistent inspiratory stridor. Lower airway anomalies like tracheomalacia, trachea-esophageal fistula, bronchomalacia, impacted foreign body, airway stenosis, granuloma, Endobronchial Tumour and thick mucus impaction may cause or exacerbate the underlying pulmonary pathology. Non-infective pulmonary causes like pulmonary hemorrhage, aspiration pneumonia, lipoid pneumonia may give characteristic features on BAL sample. Compromised airway lumen due to external occlusion by lymph nodes or vascular structures can be assessed using FB in intensive care.

Therapeutic Indications

Bronchoscopic suction of thick mucus impaction, debridement of granuloma or endobronchial tumor or retrieval of aspirated foreign body [(Figure2)] helps clear the airway lumen. Balloon dilatation can be done in cases of congenital or acquired causes of airway stenosis. Persistent bleeders from airway mucosal surface can be controlled using argon plasma coagulation (APC) and cryotherapy. FB can be used as an accessory procedure

in cases of difficult intubation, difficult extubation and selective lung ventilation. In cases with anatomical airway malformation, a self-expanding metallic stent can be deployed with the assistance of FB in operation theatre.

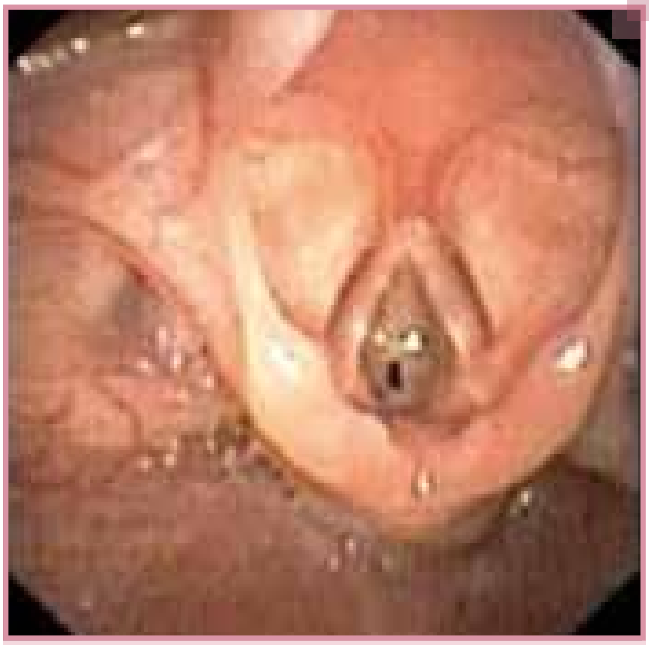

Figure 1 : Subglottic Stenosis

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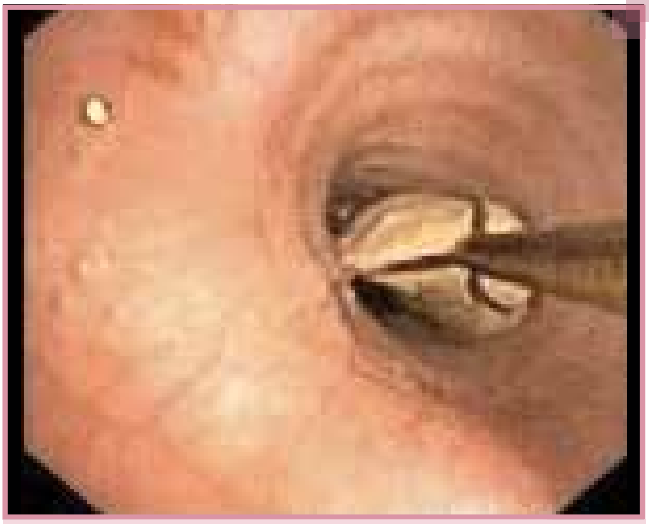

Figure 2 : Foreign Body Aspiration - Removal by Dormia Basket

Role of Computed Tomography in Pre and Postoperative Evaluation of a Double-Outlet Right Ventricle

Source: https://doi.org/10.4250/jcvi.2020.0196

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Abstract

Double-outlet right ventricle (DORV) is a type of ventriculoarterial connection in which both great arteries arise entirely or predominantly from the right ventricle. The morphology of DORV is characterized by a ventricular septal defect (location and relationship with the semilunar valve); bilateral coni and aortomitral continuity; the presence or absence of outflow tract obstruction; tricuspid-pulmonary annular distance; and associated cardiac anomalies. The surgical approach varies with the type of DORV and is based on multiple variables. Computed tomography (CT) is a robust diagnostic tool for the preoperative and postoperative assessment of DORV. Unlike echocardiography and magnetic resonance imaging (MRI), CT imaging is not limited by small acoustic window, need for anaesthesia and can be used in patients with metallic implants. Current generations CT scanners with high spatial and temporal resolution, wide detectors, high-pitch scanning mode, dose-reduction algorithms, and advanced three-dimensional post-processing tools provide a low-risk, high-quality alternative to diagnostic cardiac catheterization or MRI, and have been increasingly utilized in nearly every type of congenital heart defect, including DORV.

Keywords: Congenital heart disease; Double outlet right ventricle.

Role of CT in the Pre - and Postoperative Assessment of Conotruncal Anomalies

Source : https://doi.org/10.1148/ryct.210089

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Abstract

Conotruncal anomalies, also referred to as outflow tract anomalies, are congenital heart defects that result from abnormal septation of the great vessels' outflow tracts. The major conotruncal anomalies include tetralogy of Fallot, double-outlet right ventricle, transposition of the great arteries, truncus arteriosus, and interrupted aortic arch. Other defects, which are often components of the major anomalies, include pulmonary atresia with ventricular septal defect, pulmonary valve agenesis, aortopulmonary window, and double-outlet left ventricle. CT has emerged as a robust diagnostic tool in preoperative and postoperative assessment of various congenital heart diseases, including conotruncal anomalies. The data provided with multidetector CT imaging are useful for treatment planning and follow-up monitoring after surgery or intervention. Unlike echocardiography and MRI, CT is not limited by a small acoustic window, metallic devices, and need for sedation or anesthesia. Major advances in CT equipment, including dual-source scanners, wide-detector scanners, high-efficiency detectors, higher x-ray tube power, automatic tube current modulation, and advanced three-dimensional postprocessing, provide a low-risk, high-quality alternative to diagnostic cardiac catheterization and MRI. This review explores the various conotruncal anomalies and elucidates the role of CT imaging in their pre- and post-operative assessment.

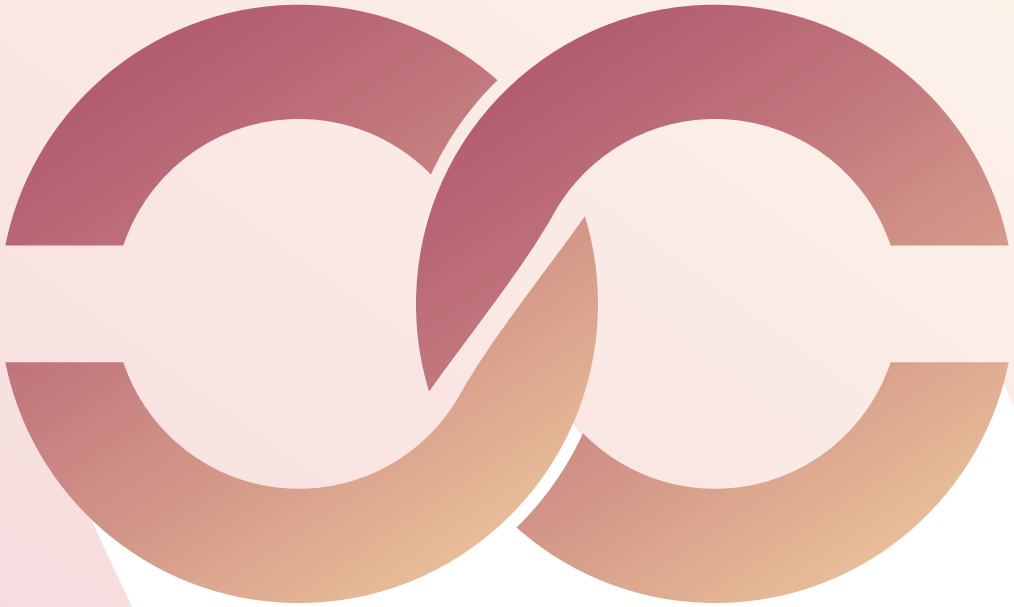
Summary

CT is a useful, non-invasive imaging modality for the pre- and post-operative assessment of conotruncal anomalies.

Essentials

The major conotruncal anomalies include tetralogy of Fallot, double-outlet right ventricle, transposition of the great arteries, truncus arteriosus, and interrupted aortic arch. nCT is a useful, non-invasive imaging modality to help assess the preoperative anatomy and postoperative complications of conotruncal anomalies. Current generations of CT scanners made with the latest tech-nologies, such as fast gantry rotation, high-pitch scanning, wider detector systems, automated tube voltage selection, tube current modulation, and three-dimensional printing, have revolutionized the role of CT in pre- and postoperative evaluation of congenital heart diseases.

Keywords: CT, CT Angiography, Stents, Paediatrics



ONCO CONNECT

Central Tumour Board: Head and Neck

Dated - 13th April, 2023

COORDINATOR	**Dr Hitesh R Singhavi** Consultant - Head and Neck, Fortis Hospital, Mulund **Dr Harshit Shah** Associate Consultant - Surgical Oncology, Fortis Hospital, Kalyan
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	MEDICAL ONCOLOGISTS **Dr Uma Dangi** Consultant - Medical Oncology, Fortis Hospital, Mulund
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Cases

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Case 1

Case Report

A 45 year old male with past history of tobacco use and ECOG-0 underwent wide excision left lateral border tongue with left neck dissection with primary closure in July 2020 (pT1N0M0). He did not undergo radiation therapy. Patient was on regular follow up. CECT showed no bony erosion and no significant cervical node appreciation. Underwent wide excision of right buccal mucosa with frozen section. HPR suggests multicentric verrucous carcinoma of right buccal mucosa with secretions of both nodular lesion and leukoplakic patch showing verrucous carcinoma at a distance of 1.2cm. Depth of invasion of nodular lesion is 0.2cm and leucoplakia patch is 0.1cm. No LVE or PNI. All margins are free of Tumor.

Discussion

Verrucous carcinoma is a subtype of squamous cell carcinoma (SCC) that typically presents as a slow-growing, well-differentiated tumour. The indication of radiation therapy in the treatment of verrucous carcinoma is very limited. Study led by Mohan et al compared overall survival of scc patients with adjuvant RT, surgery alone and verrucous carcinoma with adjuvant RT. They found that Verrucous carcinoma to have poor OS and DFS as compared to others.[1]

Decision

Neck dissection with no adjuvant treatment.

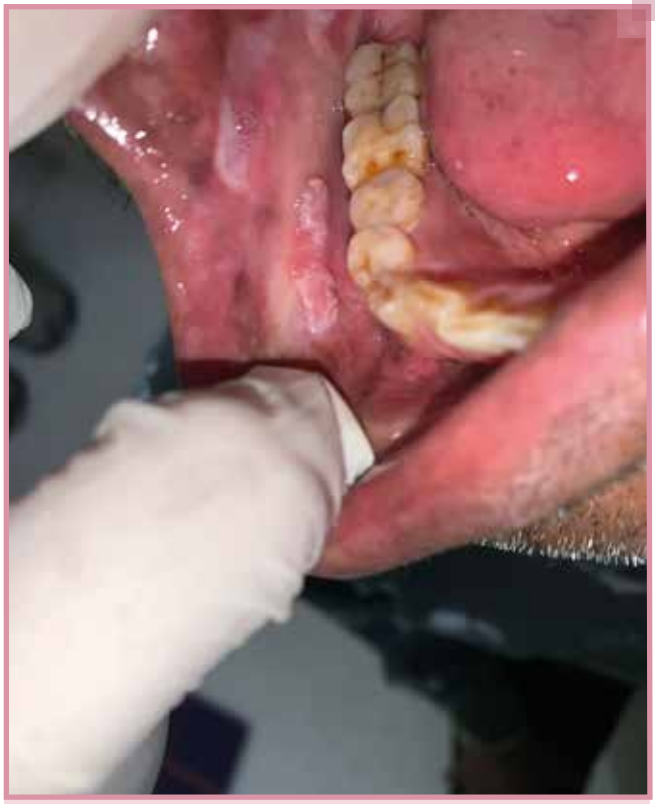

Figure 1 : February 2022 - Biopsy - Leucoplakia

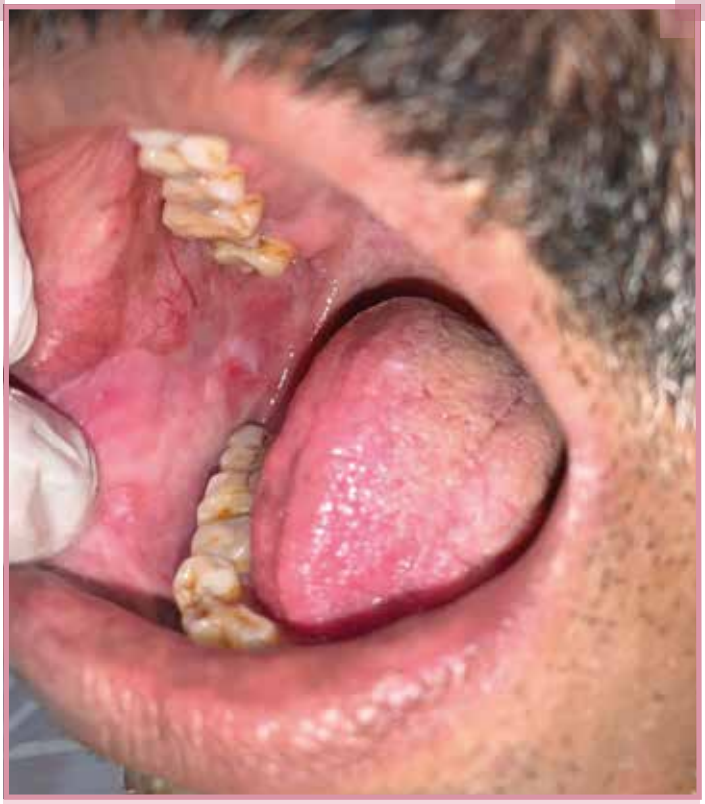

Figure 2 : February 2023- Biopsy- Verrucous carcinoma

Case 2

Case Report

A 35-year-old male, occasional alcoholic with ECOG- 0 underwent excisional biopsy of left lateral border tongue with margin positive 8 days back. HPR shows well differentiated squamous cell carcinoma. Tumor size is 1.7cm with depth of invasion of 0.5cm. LVE and PNI absent. Base is free. Two mucosal margins are involved. Indurated scar over left lateral border tongue. MRI suggests heterogeneous enhancing with few necrotic areas of size 10x25mm showing diffuse restriction with low ADC values. Underwent wide excision left lateral border tongue and left neck dissection and primary closure. HPR shows residual focus of SCC (<0.2cm). PNI and LVE absent. All nodes are reactive. All margins are clear and DOI is 2mm.

Discussion

Depth of invasion has an influence over the prognosis of the patients. DOI < 5 mm alone may not be enough to warranty post operative radiation. According to the international collaborative study done 2019 led by Ebrahmi et al stated that Depth of invasion is the factor which is associated with other adverse factor and in the absence of these factor DOI (<5mm) do not add to the factor responsible for PORT. It only adds to 4% of the total benefit in such cases.[2]

Decision

Slide review, if on slide review the depth of invasion is greater than 5mm then adjuvant radiation otherwise observe.

Case 3

Case Report

A 59-year-old male underwent wide local excision of left buccal mucosa + MND + SSG in 2016. He developed left buccal mucosa recurrence in 2020. Received NACT. Underwent left commando and ALT followed by CTRT. Then developed second primary in Palate + OAF in Jan 2021. Margins were positive. Was planned for metronomic chemotherapy. Now has ulcer on Left BM with skin involvement. Biopsy is positive for malignancy.

Discussion

PET CECT scan + triple scopy followed by surgical excision V/s Palliative Chemo.

Figure 1 : Recurrent left buccal mucosa cancers

Decision

Resection as it is a localized disease with consent of the patients and relative considering it is aggressive disease with high chances of recurrence.

Case 4

Case Report

58-year-old female with case of CA left buccal mucosa underwent left segmental mandibulectomy + Left MRND + right neck node dissection in May 2022 (T4N1). She received radiation therapy. Then underwent debridement and salvage of PMMC flap for infected FFOCF in June 2022. PET-CT findings show ill-defined enhancing soft tissue with low grade FDG uptake involving tongue. 1.8x1.8x1.0cm SUV max= 3.6. Soft tissue seen involving posterolateral aspect of reconstructed flap. 8.0x7.0mm SUV max= 4.7. Subcutaneous enhancing deposit in left cervical region overlying sternocleidomastoid muscle.1.1x0.8cm SUV max=3.1. FNAC left node III is negative for malignancy with no clinical signs and symptoms intraorally and palpable node in left neck.

Discussion

Case was discussed about the importance of clinical finding over PETCECT for oral findings and FNAC for left neck node. Negative predictive value of PETCECT is 91% according to Mac Dermot who studied more than 1500 scans which was not enough to term it as negative while two consecutive negative scan lead to NPV of more than 98%. Thus repeating the PETCECT scan was advised after 6 weeks.[3]

Decision

Observation followed by PET after 6 weeks. USG guided FNAC of the node. If node is positive, resection. If node is negative, then Repeat PETCECT after 6 weeks.

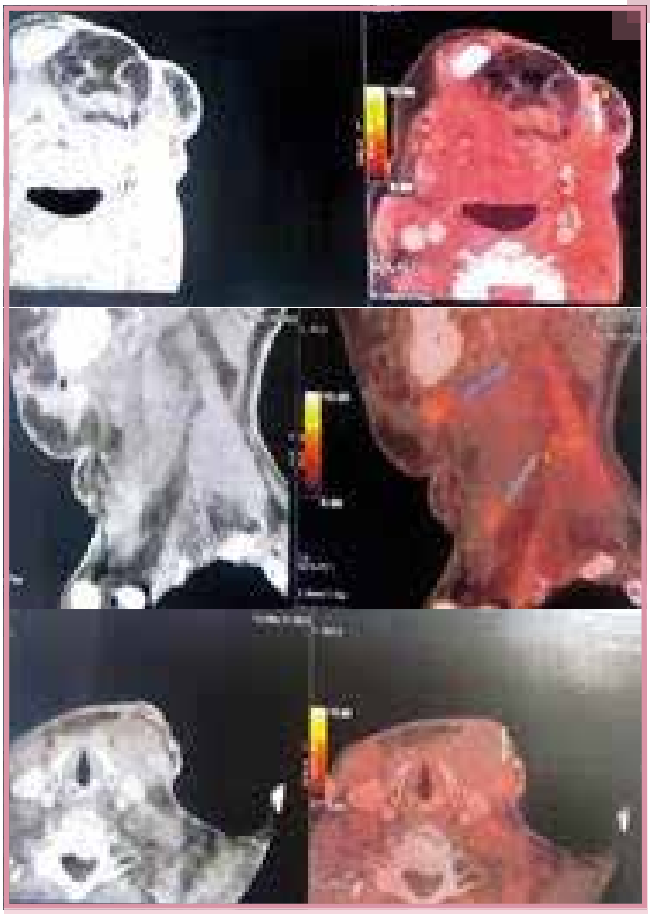

Figure 1 : FDG avid left level 1b and level 3 node

Case 5

61-year-old female, ECOG-0, No comorbidities, No habits presented with case of swelling in left base tongue with difficulty in swallowing since 3 months. Biopsy suggestive of acinic cell carcinoma, IHC showed mucoepidermoid carcinoma Intermediate grade. Fibreoptic laryngoscopy shows infiltrative lesion over left base tongue involving median gloss epiglottic fold involving left side vallecular and abutting left tonsil. Mouth opening was more than 35 mm.

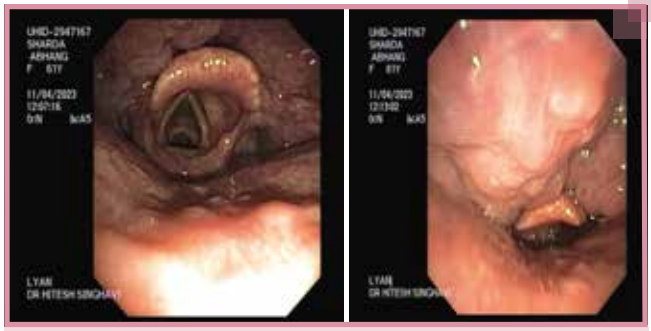

Figure 1 : Fibreoptic laryngoscopy - Infiltrative lesion over left base tongue involving median glossoepiglottic fold involving left side vallecular and abutting left tonsil

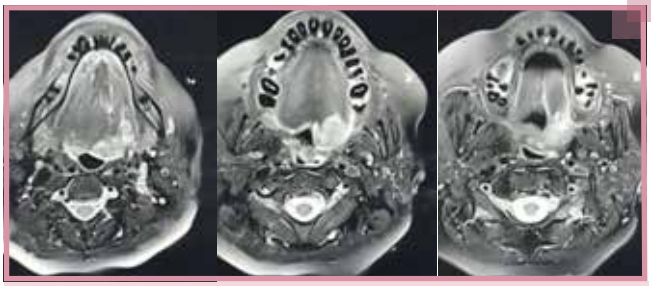

Figure 2 : MRICE - Infiltrative lesion over left base tongue involving median glossoepiglottic fold involving left side vallecular and abutting left tonsil.

Discussion

Approach to the access the lesion was discussed as Robotically assisted versus pull through and mandibulotomy approach. Since the biopsy was mucoepidermoid carcinoma and good mouth opening, Robotic approach was favored over pull through. If defect lead to communication with the neck, then reconstruction with free flap would be favorable.

Decision

Open or Robotic approach can be adopted. Robotic > open approach.

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Central Tumour Board: GI Oncology

Dated - 25th May, 2023

COORDINATOR	**Dr Ashish Sachan** Consultant – GI Surgery, Fortis Hospital, Shalimar Bagh, New Delhi
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	MEDICAL ONCOLOGISTS **Dr Ankur Bahl** Senior Director Medical Oncology, Fortis Memorial Research Institute, Gurugram **Dr Mohit Aggarwal** Additional Director - Medical Oncology, Fortis Hospital, Shalimar Bagh< New Delhi
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	ONCO-PATHOLOGY **Dr Kunal Sharma** Associate Director and Section Head Centre of Excellence and Histopathology, Mumbai Reference Labs Lead- DP and AI initiatives SRL Diagnostics

Cases

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Presenter
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Locally Advanced Desmoid Tumor Adherent to Mesenteric Vessels

Case History

A 40-year-old female with a history of two caesarean sections presented with a one-month history of abdominal pain and fullness. Physical examination revealed an intra-abdominal hard and mobile lump measuring 20cm x 13cm. Ultrasound and contrast-enhanced CT indicated an 18cm x 12cm x 15cm heterogeneously enhancing mass in the umbilical and hypogastric regions. The mass displayed soft tissue density with peripheral enhancement and notable vessel prominence. It encased and abutted at least two loops of the small bowel, causing distortion in the adjoining mesentery.

Biopsy and Imaging Findings

A biopsy of the mass showed a spindle cell neoplasm, with considerations for GIST or fibromatosis. Whole-body PET CT revealed a lobulated, heterogeneously enhancing soft tissue mass in the umbilical and hypogastric regions. The tumor abutted the anterior abdominal wall and serosal surface of small bowel loops, exhibited mild luminal narrowing, and encased distal branches of the superior mesenteric artery. Inferiorly, it approached the distal ileum and ileocecal junction. Importantly, significant nodular thickening of the adjoining mesentery was suggestive of a primary malignant disease of mesenchymal origin.

Surgical Management

The patient underwent exploratory laparotomy, during which the mesenteric tumor was resected, along with a segment of the small bowel. The tumor, measuring 18cm x 15cm, was found to be closely abutting and adherent to the superior mesenteric vessels and surrounding approximately three feet of small bowel loops. A sleeve resection of the duodenum (D4) was performed, followed by primary repair of the duodenal wall. The resected proximal and distal bowel loops were anastomosed side to side.

Postoperative Course

The patient's postoperative recovery was uneventful, and she was discharged on the 7th postoperative day. Histopathological examination confirmed the diagnosis of desmoid fibromatosis, with positive immunohistochemical staining for vimentin, SMA, and beta-catenin (nuclear staining). The Ki-67 proliferation index was 2-3%.

Points for Discussion

In this case, the tumor board was presented with several management options for this locally advanced desmoid tumor adherent to mesenteric vessels. The points discussed included the following: 1) the use of adjuvant imatinib or sorafenib, 2) close follow-up to monitor disease progression, 3) administration of sulindac and tamoxifen, and 4) the role of local radiation on the tumor along the mesenteric vessels.

Plan

The current plan is to initiate Tamoxifen therapy and closely follow up with the patient to monitor treatment response and disease progression.

Central Tumour Board: Head and Neck

Dated - 22th June, 2023

COORDINATOR	**Dr Athira Ramakrishnan** Consultant Head and Neck Oncology and reconstruction, Fortis Hospital, Bannerghatta Road, Bengaluru
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Cases

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Presenter
Dr Srivishnu S
Senior Clinical Fellow - Minimal Access Surgical Oncology,
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Case 1: Contralateral skip lesions and early neck recurrence in Carcinoma tongue.

History

- Patient is a known case of Carcinoma Tongue in the right lateral border, post wide local excision eight months back.
- Bilateral neck dissection – Right MRND (I to V) and Left level (I - IV) was done
- Post-Operative HPR revealed – pT2 N1
- Patient received adjuvant Radiotherapy – 60 Gy in 30# using IMRT technique for tumour bed and 56Gy in 30# covering bilateral neck.

Current Presentation

- The patient presented with a swelling in the left lower neck (Level V – supraclavicular region). On examination there was a firm lymph node with restricted mobility.
- PET CECT revealed an uptake of SUV 6.14 in the level V lymph node. There were no distant metastases
- Biopsy from the lymph node was suggestive of metastatic squamous cell carcinoma
- Patient had a disease-free interval (DFI) of eight months post adjuvant therapy

Points of Discussion

- **Role of Neo adjuvant chemotherapy:** In view of short DFI of 8 months and the tongue being an aggressive primary, neoadjuvant chemotherapy with cisplatin-based regimen has been planned
- **Role of Completion Neck dissection:** Following the systemic therapy, imaging to re assess the response and completion of neck dissection has been planned.
- **Role of re-irradiation:** Re irradiation will be considered based on the response. A lesser dose of 20 -30Gy will be planned

Case 2: Fungating Right parotid mass with doubtful resectability.

History

- 45 years female, with history of progressively growing right parotid mass for 12 months. It was associated with loss of facial nerve function and involvement of the mandible. Core Biopsy - histopathology report showed poorly differentiated carcinoma with features suggestive of myoepithelial carcinoma.
- PET CECT showed loco regional disease with involvement of the bilateral lymph nodes. No distant metastasis was found. The Right Internal jugular vein was infiltrated for > 360 degrees from the jugular foramen to the **level III** neck region. The common and the internal carotid artery was free. The branches of the external carotid artery, including the facial and superior thyroid arteries were involved. The plane with the mastoid process was obliterated and the posteriorly the upper portion of the trapezius was involved.
- The patient was given the option of trial resection versus neo adjuvant chemotherapy. The patient opted the latter.

Neoadjuvant Chemotherapy (NACT)

- The patient received three cycles of Paclitaxel and carboplatin, followed by three more cycles with added transtuzumab.
- Immunotherapy with pembrolizumab was started for the last two cycles as a bridge to surgical resection.

Response Assessment

- In the response assessment using PET CECT, patient did not have any distant failures. The primary had partial to stable response, with similar vascular relations seen in the pre-chemotherapy scan.
- Patient was re counselled for trial resection and the patient consented for the same.

Surgery

- Patient underwent Wide local composite resection with Right radical parotidectomy with facial nerve resection at the stylomastoid foramen entry point with hemi mandibulectomy, Right Internal jugular vein ligation at the jugular foramen, right Exteranal auditary canal (EAC) resection and Mastoid process drilling; Right Radical neck dissection and left Modified radical neck dissection and Right Tarsorraphy followed by Free Antero Lateral thigh flap (FALT) reconstruction.

Histpathology

- Residual Poorly differentiated carcinoma post NACT
- IHC: Positive for Pan CK, CK 7, Ki 67 90% and Her 2 status was negative
- Suggestive of high-grade Myoepithelial carcinoma
- 14 x 10 x 11cm size
- All margins including the EAC, Mastoid region, mandibular bony section and facial nerve were free
- Ipsilateral 5/ 17 lymph nodes were positive. Contralateral nodes were free of tumour.
- Perineural Invasion positive; No ENE

Points for Discussion

- Adjuvant radiotherapy:

After discussion in the multi-disciplinary tumour board, Adjuvant radiotherapy was planned for the patient. Dosage of 60 Gy in 30# to post op bed and right neck; 54 Gy in 30# to opposite neck using VMAT technique will be optimal.

Case 3: Role of completion neck dissection in post-operative incidental diagnosis of medullary carcinoma of thyroid

History

- 56 years male patient is a presented with complaints of swelling in the left lateral neck since 6 months
- USG neck revealed TIRADS 5 lesion in the left lobe thyroid
- USG neck further showed enlarged left level II lymph nodes largest being 2x2cm
- FNAC of the thyroid nodule was suggestive of papillary carcinoma thyroid (done elsewhere and not reviewed).
- FNAC of the cervical lymph node showed metastatic carcinoma.

Figure 1 : Pre-operative – Fungating right parotid mass (A-B) demonstrating the right facial nerve palsy (C-D)

Figure 2 : Post-operative - Following the FALT reconstruction and right tarsorrhaphy.

Surgery

- Patient underwent Robotic total thyroidectomy with left central compartment node dissection and left lateral neck dissection(level II to V)

Histopathology Report

- Medullary carcinoma thyroid
- Positive for Congo red stain
- Left lateral neck node positive for metastasis.

Points for Discussion

- Role of Tumour markers : Patient's Serum Calcitonin and CEA levels three weeks after surgery were 64 and 4 respectively. This would be kept as a baseline for further follow up.
- Role of Genetic testing: Patient underwent germline genetic testing. It showed a mutation in the exon number 618 which belonged to the moderate risk category.
- Role of further imaging: Patient underwent a DOTA NOC PET which revealed no local or distant disease in the body
- Role of Completion Neck dissection: In view of the Calcitonin level less than 150, no residual disease elsewhere in the body and a moderate risk mutated codon, it was in the multidisciplinary tumour board to observe and follow up the patient with Serum calcitonin and CEA levels combined with USG of the neck on the three-monthly basis for the first three years.

Case 4 : Case of Osteoradionecrosis Fibula

Presenter
Dr Hitesh R Singhavi
Consultant – Head and Neck Surgery,
Fortis Hospital, Mulund, Mumbai

Case report

48-year Male, complained of plate exposure and foul-smelling pus discharge from lower alveolus since 2months. He is an operated case of extended middle third mandibulectomy with bilateral neck dissection with free fibula in May 2020. (Three year back). Biopsy from the suspicious mucosal and bony lesion suggested of osteonecrosis of fibula. There were no neck nodes palpable or on scans. PETCECT was done which showed to no suspicious uptake anywhere in the body except at the lower right reconstructed fibula. Reconstructed fibula showed mild non-FDG uptake features suggestive of graft resorption and non-union with secondary infection.

Discussion

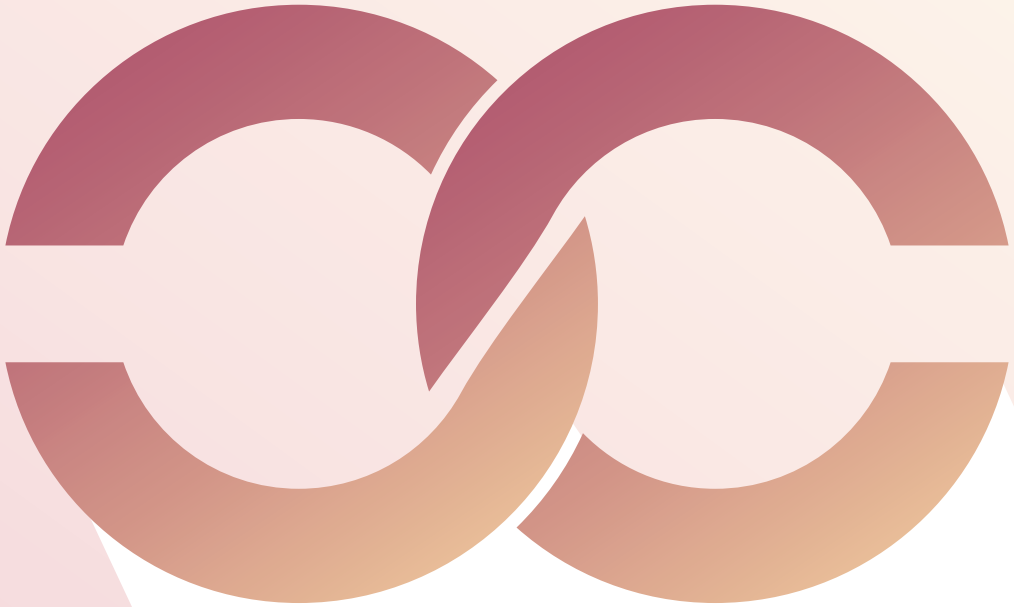
Case was discussed for options of reconstruction involving removing plate with fibula and reconstruction with another fibula, removal of plate and fibula and reconstruction with PMMC and DP. Removal of plate with removal of infected segment and constructing with PMMC/DP. Considering the patient has undergone surgery and radiotherapy in past option of reconstruction with fibula and another flap for covering the outer defect was ruled due to the extensiveness of the surgery. Reconstructing the defect with only soft tissue flap would hinder his ability and access to nutrition per orally and he might be ryles tube dependent for long time.

Decision

Unanimous decision was to remove the screws with infected segment and cover it with PMMC. To counsel the patient regarding the chances of non-healing wound leading to requirement of another surgery.

Figure 1 : Mandibular fibular Plate exposure after 2 years of surgery

Figure 2 : Malunion of osteotomy of fibula



CLINICAL RESEARCH

Targeted Individualized Versus Standardized Preterm Human Breast Milk Fortification: A Randomized Controlled Trial

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Introduction

Extrauterine growth retardation (EUGR) is one of the major problems faced in most preterm neonates. The only way to prevent EUGR is by providing adequate number of calories through fortification of human breast milk. Human milk fortification (HMF) is now considered a standard practice to prevent EUGR in most of the neonatal units. We compared targeted individualized fortification (TIF) versus standardized fortification (SF) of breast milk with HMF to assess catch-up growth in preterm neonates. cv

Keywords: Fortification, HMF, macronutrients, milk analysis, preterm nutrition Targeted Individualized versus Standardized Preterm Human Breast Milk Fortification: A Randomized Controlled Trial

Materials and Methods

This single-centre prospective randomized control study was conducted at a tertiary level NICU in New Delhi, India. This study was performed according to the principles of the Declaration of Helsinki. Ethics committee approval was taken from the institutional ethics committee; the study was also registered with Clinical Trials Registry – India (CTRI/2018/05/014195). Informed consent was taken from the parents of all the neonates enrolled into the study.

Randomization

Neonates ≤32 weeks of gestation or those with birth weight ≤1800 g were randomized into two groups: targeted individualized fortification (TIF) group and standard fortification group (SF) with a blinded draw method; the allocation ratio of the two groups during the draw was always 1:1. The randomization sequence

Outcomes

The primary outcome of the study was to compare the TIF with standard fortification of human breast milk with HMF in preterm neonates to assess the catch-up growth between the two groups, both in terms of weight and head circumference. Catch-up growth was defined as neonates achieving their birth centile of weight and head circumference according to the Fenton's growth chart.

Results

Thirty-two neonates were enrolled into the study and randomized – 15 to the TIF group and 17 to the SF group [Figure 1]. Seven neonates (TIF: n = 2; SF: n = 3) were excluded from the study analysis due to switch to formula, deviation from feeding protocol (nil per orally), or death.

The two groups were comparable in weight, gestation, and head circumference at the time of enrolment into the study The mean gestation age at birth was 29.4 weeks (±2.78) in the TIF group and 29 weeks (±2.33) in the SF group. The mean birth weight and head circumference were 1268.76 g (±315.66) and 27.23 cm (±2.94) in the TIF group and 1172.83 g (±331.95) and 26.58 cm (±2.4) in the standard group, respectively (both P > 0.05). Parenteral nutrition and subsequent enteral feeding were started and given similarly in both the groups, as per protocol. There was a significant increase in the mean birth weight and mean head circumference of the neonates in the TIF group versus

the SF group (P = 0.00071 and P = 0.00175, respectively) at the end of the study period.

Conclusion

TIF is feasible in clinical practice and is better for achieving catch-up growth in preterm neonates without any adverse effects. Targeted fortification helps mitigate some of the drawbacks of standard fortification of human breast milk for preterm nutrition. TIF is a potential way forward to prevent/decrease EUGR.

Financial Support and Sponsorship

Nil.

Conflicts of Interest

There are no conflicts of interest.

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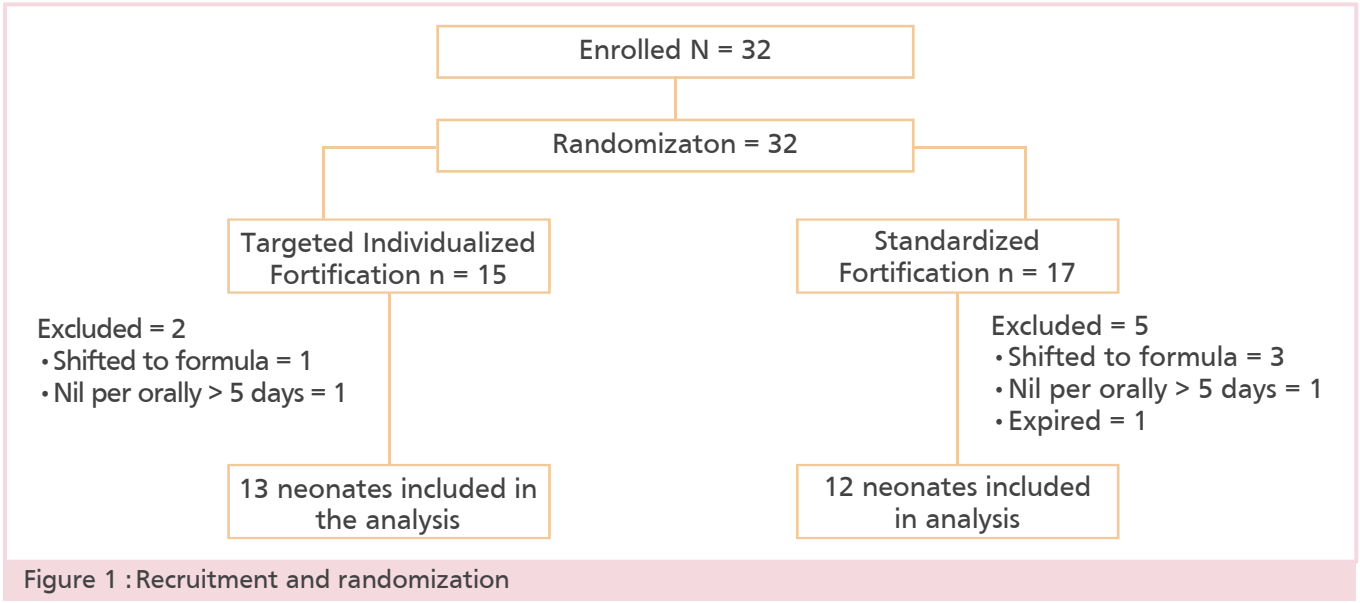
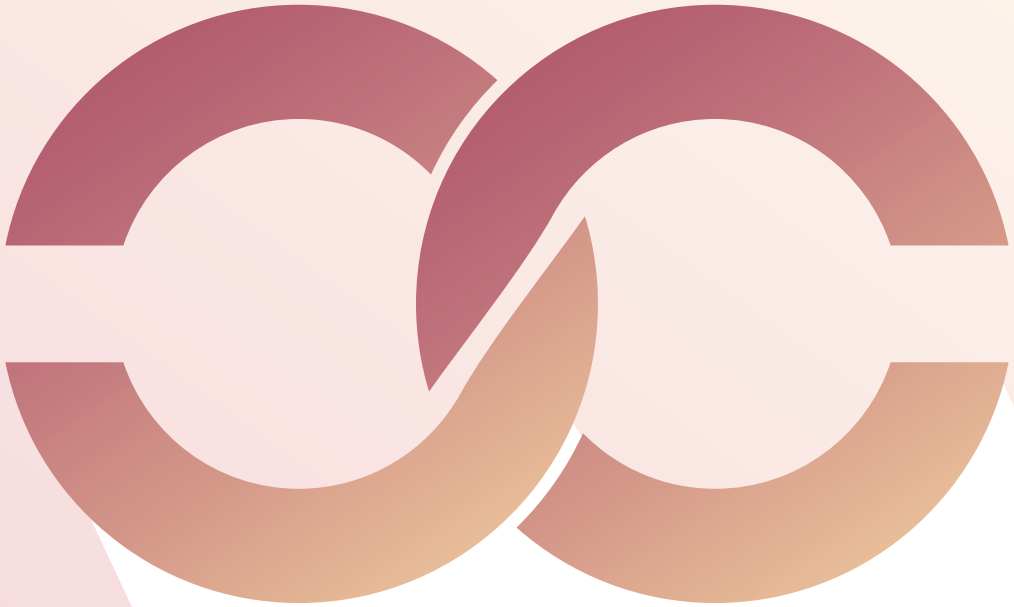


Figure 1 : Recruitment and randomization



MEDICATION SAFETY UPDATE

MEDICATION SAFETY UPDATE

U.S.Food and Drug Administration (FDA) approved JARDIANCE (EMPAGLIFLOZIN) and SYNJARDY (EMPAGLIFLOZIN AND METFORMIN HYDROCHLORIDE) as Additions to Diet and Exercise to Improve Blood Sugar Control in Children 10 Years and Older with Type 2 Diabetes.

Source: www.fda.gov, June 20, 2023

This approval provides much-needed additional treatment options for children with type 2 diabetes as Compared to adults, children with type 2 diabetes have limited treatment options, even though the disease and symptom onset generally progress more rapidly in children.

Metformin, the only other oral therapy available for the treatment of children with type 2 diabetes, was first approved for paediatric use in 2000.

EMPAGLIFLOZIN, the active ingredient in Jardiance and Synjardy, works by increasing the excretion of glucose in the urine.

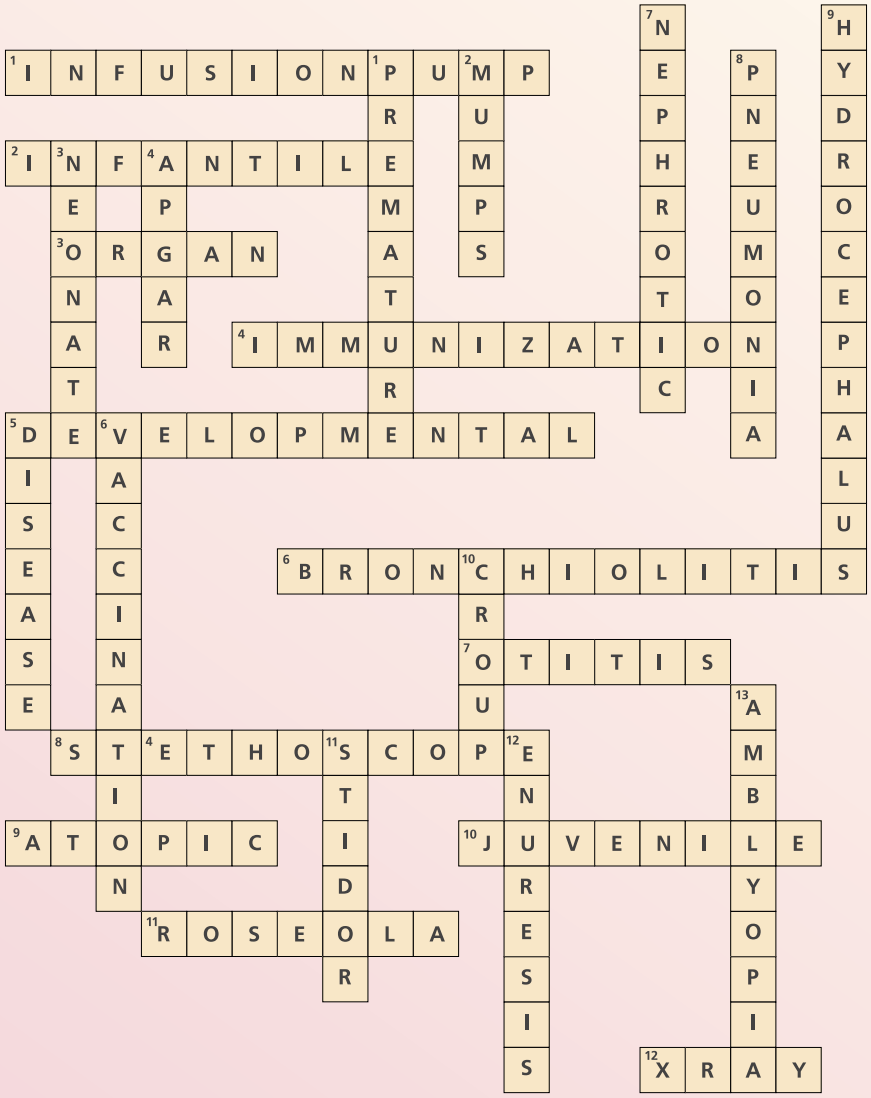
The Common Side Effect of EMPAGLIFLOZIN in Children

- **Ketoacidosis (increased ketones in your blood or urine).** Ketoacidosis is a serious condition that needs to be treated in the hospital. **Even if blood sugar is less than 250 mg/dL.** Ketoacidosis has also happened in people with diabetes who were sick or who had surgery during treatment with EMPAGLIFLOZIN, and if possible, check for ketones in your urine: nausea, vomiting, stomach-area (abdominal) pain, tiredness, or trouble breathing.
- **Dehydration.**
- **Serious urinary tract infections.**
- **Low blood sugar (hypoglycemia).** Symptoms of low blood sugar may include headache, drowsiness, weakness, dizziness, confusion, irritability, hunger, fast heartbeat, sweating, shaking or feeling jittery.
- **Necrotizing fasciitis.** A rare but serious bacterial infection that causes damage to the tissue under the skin in the area between and around your anus and genitals (perineum). This bacterial infection has happened in women and men who take EMPAGLIFLOZIN.
- **Vaginal yeast infection.**
- **Yeast infection of the penis**
- **Allergic (hypersensitivity) reactions.**

Contraindications

- Patients with **Type 1 diabetes**
- Patients with **Severe kidney problems**
- Patients who previously have had a serious **Allergic reaction** to EMPAGLIFLOZIN
- Patients with **Metabolic** acidosis or **Diabetic ketoacidosis**

Answer To The Crossword



Across

1. Infusion Pump
2. Infantile
3. Organ
4. Immunization
5. Developmental
6. Bronchiolitis
7. Otitis
8. Stethoscope
9. Atopic
10. Juvenile
11. Roseola
12. Xray

Down

1. Premature
2. Mumps
3. Neonate
4. Apgar
5. Disease
6. Vaccination
7. Nephrotic
8. Pneumonia
9. Hydrocephalus
10. Croup
11. Stidor
12. Enuresis
13. Amblyopia

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