



News Letter

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Too often we underestimate the power of a touch, a smile, a kind word, a listening ear, an honest compliment, or the smallest act of caring, all of which have the potential to turn a life around.

...Kahlil Gibran



Case of the month :
Unusual treatment of a movement disorder

From the Managing Director's desk



- Teams are not ends in themselves; they
- are a means by which to achieve other
- organizational goals.

Richard Wellins

Dear Colleagues,

As you all know we have expanded our services in a major way in the last one year. Now with involvement in '**Rajiv Aarogyasri**', a unique health insurance scheme, is being implemented efficiently and successfully in Andhra Pradesh we are able to provide the best quality of care to poor and needy.

White cardholders and their family members can avail themselves of the scheme, which facilitates 272 types of surgeries including those for congenital defects.

An '**Aarogyamitra**' at the hospital will attend to the patient right from admission to discharge after surgery. Those not eligible for benefits under '**Aarogyasri**' can seek assistance from the Chief Minister's Relief Fund.

Rainbow Hospitals is the largest group of children's hospitals with 275 beds – 120 of them devoted exclusively for intensive care at Banjara Hills, Hyderabad, Secunderabad, and Vijayawada.

Rainbow has a 10-year track record of excellence in handling all types of pediatric disorders thanks to the expertise and experience of its medical and support personnel, perfect teamwork and state-of-the-art operation theaters. Rainbow has the largest multidisciplinary intensive care unit to support any complex surgical or medical **problem**. Barring cardiac surgeries, Rainbow performs the one of largest number of neo-natal, liver, gastro-intestinal, renal, orthopedic and neuro surgeries in children.

Dr. Ramesh Kancherla

Surgeries covered under

'Aarogyasri'

Neonatal Surgery

Esophageal Atresia
Congenital Diaphragmatic Hernia
Neonatal Intestinal Obstructions
Anorectal Malformations
Congenital Mega Colons

Pediatric Urology

Congenital Hydronephrosis
Hypospadias
Wilm's Tumor
Exstrophy of Bladder
Ureteric Reimplantations
Posterior Urethral Valves

Pediatric Neurosurgery

Pediatric Hepatobiliary Surgery

Biliary Atresia
Choledochal Cyst
Liver Tumors
Meningocele
Meningo myelocele
Congenital Hydrocephalus

Pediatric Plastic Surgery

Cleft Lip
Cleft Palate
Syndactyly
Post Burns Contracture

Pediatric Surgical Oncology

All pediatric Tumors

Syndactyly

Post Burns Contracture

Pediatric Surgical Oncology

All pediatric Tumors



From the Editor's Desk

Dear Colleagues,

There had been a long hiatus between the last RCH Newsletter and the current one. Hence we are starting afresh. The format will contain few reviews of common Paediatric Topics, 1-2 interesting cases of the month. Henceforth the Newsletter will be published thrice in a year.

There have been exciting new development at RCH during this period. We have expanded with branches in Vikrampuri, Secunderabad and Gandhinagar at Vijayawada. They are fully equipped with Neonatal and Paediatric Intensive Care services, with visits from various Paediatric subspecialists at regular intervals for outpatient services. The details are available on the website (www.rainbowhospitals.in).

We have had expansion in terms of Paediatric subspecialists as well during the intervening period which many of you are already aware. To quickly introduce

- 1) **Dr Satish Ghanta- Consultant Paediatric Intensivist and Neonatologist** with training from Prestigious units at Australia. He is a very gentle and well mannered Paediatrician.
- 2) **Dr Aparna Reddy – Consultant Paediatrician and Pulmonologist**, with Paediatric Pulmonology training at Singapore. She has expertise in bronchoscopy as well.
- 3) **Dr Sirisha S- Consultant Paediatric Haemato Oncology and BMT**. Her Haem Onc training started in Kidwai Memorial hospital in Bangalore with further training in Prestigious units at UK including GOSH, UCL and Bristol Children's Hospital
- 4) **Dr Lokesh Lingappa- Consultant Paediatric Neurologist**. After DM Neurology from Bangalore, did Fellowship in Paediatric Neurology at UK.
- 5) **Dr Radha Ramadevi- Senior Consultant Paediatrician with expertise in Genetics and metabolic disorder** is a familiar name to many Paediatricians within the state and also in the Country. She is going to joins us soon and will be heading the Genetic and Metabolic services.

To sign off I will be handing over the editorial responsibilities to Dr Lokesh who is a keen teacher and is interested in carrying on this activities for now. Wishing him all the Best. Enjoy the reading.

Your's Sincerely

Dr Mehul Shah

Consultant Paediatric Nephrologist.
MD (Peds), DCH, AB Peds (USA),
AB Ped.Nephrology (USA)

Meet Our Contributors



Dr.A.Radha Rama Devi

Doctor of Medicine from Andhra Medical College, Specializing in the field of Pediatrics. Certificate course in Genetic Engineering from Indian Institute of Science, Bangalore.

ACADEMIC CREDENTIALS:

Awarded merit prizes in MBBS including the President's Medal and Gold Medal in Medicine. She did training fellowship by British Council in the field of Inborn Errors of Metabolism Senior Women Bioscientist in India for the year 2005 for life time achievement by the Department of Biotechnology, Government of India.

Career Achievements:

40 years of practice in clinical Pediatrics and research and in the area of Medical Genetics. Set up the Nation's comprehensive Genetic Diagnostic Service Laboratory for the diagnosis of Biochemical, molecular and cytogenetic disorders at Center for DNA Fingerprinting and Diagnostics, a unit of Department of Biotechnology Government of India. This led to the setting up of prenatal diagnosis for the high risk families with the objective of genetic counseling and prevention of genetic disorders.

Instrumental in setting up the first Newborn Screening for preventable causes of Mental retardation in India and published the first data on the prevalence of these disorders in the Indian population. This led to the starting of pilot screening by Indian Council of Medical Research for the most common disorders in five centers in the country now.

Dr.Lokesh

is working as fulltime Pediatric Neurologist at Rainbow Children's Hospital. He did his MBBS from KMC, Hubli. Pediatric training was at PGIMER, Chandigarh.

During his registrar training at PGI realized the need of Pediatric Neurology Services and joined DM Neurology programme at NIMHANS, Bangalore. He was awarded Anisya Vasanth Memorial award for Best Neurology Resident for year 2003. To pursue further training in Pediatric Neurology went to UK. He worked as Registrar in Pediatric Neurology at Birmingham Children's Hospital, Bristol Children's Hospital and Frenchay Hospital. He has several international presentation/ posters/papers to his credit.

To fulfill the current unmet needs of Paediatric Neurology patients in India he returned in August 2007. Areas of interest Infantile seizures, Neurometabolic disorders and Preventive Paediatric Neurology.



Dr.Sirisha

is working as a Paediatric Hemato Oncology Consultant at RCH since Aug 2007. She is an efficient and caring professional. She did her MBBS from Kurnool Medical College followed by MD Pediatrics at PGIMER, Chandigarh, She was awarded Best outgoing student. She has done her DNB paediatrics. Developed immense interest in Pediatric Haematology Oncology and worked in Kidwai Oncology institute, with Dr.Appaji Bangalore. She went to UK for further Pediatric Hemato Oncology training and worked there for 3 years in places like Great Ormond Street Hospital, Bristol Children's Hospital and

University College Hospital, London. Along with Pediatric Hemato Oncology, she also gained experience in Pediatric Bone Marrow Transplantation.

P Aparna Reddy

Completed MBBS from MR Medical college Gulbarga in the year 1994. MD Pediatrics from Dr B.R.A.M.C.,Bangalore in the year 1998.

Fellowship in pediatric pulmonology from National University Hospital,Singapore in the year 2006.Trained in pediatric pulmonology and sleep medicine.Good experience in pediatric bronchoscopy.

Area of Interest Asthma diagnosis and management.Presently managing asthma clinic equipped with state of the art exercise challenge test and pulmonary function tests.





Diagnosis and Management of Inborn Errors of Metabolism



Dr. A.Radha Rama Devi,

MD Ped, Cert. in Genetic Eng., Fellow in IEM (UK)

Senior Consultant Pediatrician Incharge Genetic Division

Improvements in medical technology and knowledge of the human genome are resulting in significant changes in the diagnosis, classification, and treatment of inherited metabolic disorders. As a result, many known inborn errors of metabolism are detected earlier and treated. It is important for primary care physicians to recognize the clinical signs of inborn errors of metabolism and to know when to pursue advanced laboratory testing or referral to a children's subspecialty center.

Inborn errors of metabolism (IEM) though individually rare but collectively are common and can present at any age even in adulthood. Diagnosis does not require extensive knowledge of biochemical pathways. An understanding of the broad clinical manifestations of IEMs provides the basis for knowing when to consider the diagnosis. A high index of suspicion is most important in making the diagnosis. Prompt therapy and emergency treatment leads to successful metabolic stabilization.

What are Inherited Metabolic Disorders?

Inborn Errors of Metabolism are inherited disorders due to deficiency of specific enzymes involved in the metabolism of proteins, carbohydrates and fats leading to a metabolic block. The symptoms are due to toxic accumulations of substrates before the block, intermediates from alternative metabolic pathways, defects in energy production and deficiency of end products beyond the block. Nearly every metabolic disease has several forms with varying age of onset, clinical severity, and, mode of inheritance.

The categories of IEMs include disorders of protein metabolism (Amino acid disorders, organic acid disorders, urea cycle disorders); Disorders of carbohydrate metabolism (e.g., carbohydrate intolerance disorders, glycogen storage disorders, disorders of gluconeogenesis and glycogenolysis) Lysosomal storage disorders, Fatty acid oxidation defects, mitochondrial disorders, Peroxisomal disorders.

IEMs can affect any organ system and usually affect multiple organ systems. Manifestations vary from those of acute life-threatening disease presenting as metabolic emergency to sub acute progressive degenerative disorder.

When to suspect an IEM?

It is very essential to consider an IEM in any critically ill neonate especially when there is a history of deterioration after an initial period of apparent good health in a term infant. A negative newborn screen does not exclude diagnosis of metabolic disease. Metabolic disease should be considered in infants with hepatoencephalopathy, poor feeding, failure to thrive, and developmental delay and in older children (>5 y), adolescents, or adults, metabolic disease should be considered with subtle neurological or psychiatric abnormalities. Onset of symptoms with change in diet and unusual dietary preferences, particularly protein or carbohydrate aversion, decompensation out of proportion to intercurrent infection, unexplained neonatal or sudden infant deaths in siblings, metabolic disease should be suspected. A negative family history does not rule out IEM. Consanguinity increases the likelihood of autosomal recessive disorders.



Diagnosis and Management of Inborn Errors of Metabolism

Clinical Presentation:

IEMs of energy deficiency, symptoms usually develop within 24 hours of birth. Neonates with inborn errors that result in defects in energy production and use often have dysmorphic features, skeletal malformations, cardiopulmonary compromise, organomegaly, and severe generalized hypotonia. Certain metabolic diseases may be associated with an increased risk of sepsis, for e.g. galactosemia, organic acidopathies, and congenital adrenal hyperplasia. Dysmorphic or coarse features, skeletal abnormalities, and abnormalities of the hair or skin, poor feeding, failure to thrive, dilated or hypertrophic cardiomyopathy, hepatomegaly, jaundice, and liver dysfunction, developmental delay, ataxia, hypotonia or hypertonia, and visual and auditory disturbances could be associated with a metabolic disease. Clinical findings in older children, adolescents, and adults include mild-to-profound mental retardation, autism, learning disorders, behavioral disturbances, hallucinations, delirium, aggressiveness, agitation, anxiety, panic attacks, seizures, dizziness, ataxia, exercise intolerance, muscle weakness, and paraparesis. Some manifestations may be intermittent, precipitated by the stress of illness, or progressive, with worsening over time.

Basic Laboratory Data:

Initial laboratory evaluation

Complete blood count (CBC) for the presence of neutropenia, anemia, and thrombocytopenia.

Serum electrolytes, bicarbonate, and blood gases for electrolyte imbalances and evaluate anion gap and acid/base status.

Blood urea nitrogen and creatinine levels to evaluate renal function.

Liver function tests, prothrombin time, and activated partial thromboplastin time to evaluate hepatic function.

It is essential to test for hypoglycemia and hyperammonemia especially when there is ketosis and acidosis. Determine the urine pH, ketone bodies and reducing substances in the urine to detect disorders of carbohydrate metabolism.

Ammonia: Is done on arterial blood preferably in the presence of altered level of consciousness, persistent or recurrent vomiting, primary metabolic acidosis with increased anion gap, or primary respiratory alkalosis in the absence of toxic ingestion. If a venous sample is obtained, the sample must be free flowing without a tourniquet, collected on ice and analyzed immediately. Normal values are <100 mcg/dL in the neonate and <80 mcg/dL in those older than 1 month.

Blood Lactate is an important investigation to rule out hypoxia and may be the cause of metabolic acidosis and mitochondrial disease.

The basic investigations primarily help in considering the presence of a metabolic disorder.

Other Tests:

Histopathology of affected tissues such as skin, liver, brain, heart, kidney, and skeletal muscle. Imaging and radiological survey, Immuno-histochemistry when required.

Specific Tests:

Based on the initial test results, specific diagnostic tests like quantitative amino acid assays, organic acids by GC-MS, Acylcarnitine profile, orotic acid assay, vitamin B12 assay etc are important for a confirmatory diagnosis.

Confirmation can be done by Enzyme assay or DNA analysis in leukocytes, erythrocytes, skin fibroblasts, liver, or other tissues.

Cerebrospinal fluid (CSF) analysis for lactate, pyruvate, organic acids, neurotransmitters, and/or disease-specific metabolites should be collected at the same time as plasma (1-2 mL)



Diagnosis and Management of Inborn Errors of Metabolism

Postmortem diagnosis:

If a child has died, attempting to diagnose a metabolic disease is still important because of the possibility that asymptomatic siblings may be affected or for the purpose of genetic counseling. Plasma, serum, urine, and possibly CSF, skin, and selected organ specimens should be collected and frozen for any future studies.

Management of IEM:

Initial management is focused on the type of clinical presentation. Acute metabolic crisis should be managed with proper calorie maintenance. Correction of hypoglycemia, electrolyte imbalance and hyperammonemia is extremely essential.

Hemo/peritoneal dialysis is indicated to remove the excess toxic metabolites.

Once metabolic compensation is achieved, specific therapies are initiated either with special diet, megavitamin therapy, enzyme replacement or chelating agents etc.

Follow-up:

It is utmost important to have a good follow-up with metabolic testing to ensure that the patient is metabolically stable.

Conclusion:

Majority of IEM's are treatable provided an early diagnosis is made. Early detection and intervention helps in preventing a child from becoming mentally retarded or blind, deaf or before the child dies in metabolic keto-acidosis. Some of the IEM's are now being detected by Newborn Screening before the development of acute signs of decompensation so as to initiate early intervention and prevent the sequelae of IEM.

Healthy Tips

- Make sure you drink plenty of water. We should drink 3-4 liters per day depending on body weight.
- You should always stretch before you work-out to avoid injury.
- Try eating smaller portions more of the throughout the day. (4-5 times per day)
- Remember a portion is the size of the palm of your hand.

Follow these simple guidelines and you will start seeing results in no time!





Dr Lokesh Lingappa,

MD Paed PGI, DM Neurology, MRCPCH, Fellow in Paediatric Neurology (UK)
Consultant Paediatric Neurologist



Infantile seizures are by definition onset of seizures between 1 month of age to two years of age. The etiology of seizures are varied in this age group as compared to seizures at any other age groups and often difficult to identify the exact semiological classification of epilepsy as well. This review will focus on the most common causes of infantile seizures and their management in brief.

Febrile Seizures

The information on febrile seizure contains statistics which is useful when considering various options in management and counselling the families.

Febrile seizures is the most common seizure disorder with peak age of onset being 18-22 months ranging between 6 months to 6 years. Prevalence being 3% with an incidence of 460/100000.

Eighty percent will have seizures within 24 hour of fever onset, GTCS being most common.

Other seizure types include myoclonic jerks in a small group of children.

Simple FS is seen in 70%, the duration being 3 to 6min in 92%(<15min)

Post ictal stage will not be more than drowsiness.

Any child with significant neurological impairment, lateralizing deficit after presumed febrile seizure will have to be investigated for alternative pathology.

Recurrence - Half of them will have recurrence, 32% have one, 15% have two and 7% will have three or more seizures.

Who are at risk of recurrence- First FS occurring in first year of life, FS occurring during low-grade febrile illness, family history of FS in first degree relative, there are persistent neurological abnormalities and first FS is complex in nature.

Risk of FS for younger siblings is 10-20%, Monozygotic twins- 70% concordance

EEG- no specific changes and is not advocated (AAP recommendation)

Prognosis - for future afebrile seizures/epilepsy

Simple febrile seizures- twofold excess(1%)

Complex febrile seizures- 6-45% risk of further seizures depending on number of complex factors(Prolonged seizure, multiple episodes, focal seizures)

Role of Clobazam during fever for intermittent prophylaxis. An RCT found that clobazam given during a febrile episode significantly reduced the rate of seizure recurrence compared with placebo (6/48 [12%] episodes with placebo v 1/60 [2%] episodes with clobazam; P = 0.01). Overall this is a cost effective measure if it can prevent seizure recurrence and its attendant hospitalization charges. Clobazam has to be given for 2 days as more than 95% of typical FS are noted within first 48 hours of onset of fever.

Prophylaxis not routinely indicated, but considered when there are 1) complex febrile seizures, 2) Neurological abnormalities 3) frequent recurrences. In presence of above features options can be discussed with the family and if agreed by parents after discussing the pros and cons can be started. The prophylaxis will not alter the future risk of afebrile seizures or epilepsy.

The best medication for prophylaxis is Phenobarbitone, Sodium Valproate may have to be avoided less than 2 years of age because of potential hepatotoxicity and pancreatitis. Carbamazepine and Phenytoin are not useful for febrile seizure prophylaxis.



Infantile Seizures- Review

Generalised epilepsy with febrile seizures plus (GEFS+)

Usually presents with Onset between 1st month to childhood, can present with initial typical febrile seizure but later can develop multiple seizure types. These include afebrile seizures, myoclonic jerks, atonic, absences and complex febrile seizures. This condition is inherited in autosomal dominant fashion with genetic heterogeneity. There are quite a few families in Andhra Pradesh with this characteristic syndrome when a detailed family history is taken in children presenting with febrile seizures. GEFS+ is shown to be secondary to mutations in SCN1A, SCN1B and SCN2A genes on Chromosome 19q. A small percentage of these families have mutations on GABRG2-GABA_A-receptor gamma 2 subunit on chromosome 2q.

EEG changes range from normal to occasional spike wave discharges.
Overall Prognosis- usually benign and self limited. Most remit by 11 years of age
If recurrent febrile seizure or other types of seizures are noted- to start on Valproate as the initial medication.

Dravet syndrome (Severe myoclonic epilepsy of Infancy)

Dravet syndrome is a rare progressive encephalopathy with Onset < 1 year. Incidence approximately is 1/30,000. Tetrad of early onset febrile seizures, myoclonic jerks, atypical absences, complex focal seizures. Initially focal febrile seizures are seen, less often bilateral seizures are noted. One fourth- progress to convulsive status
Later myoclonic seizures, segmental/generalized develop. Complex focal seizures are noted in 50%

The disease process evolves and is described in three periods

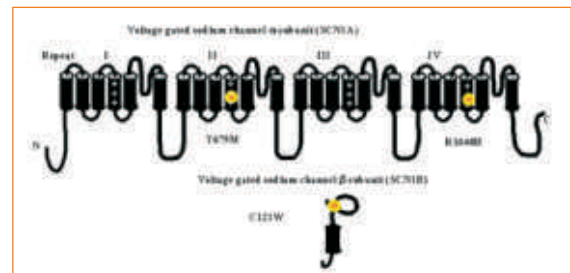
Pre-Seismic period - febrile convulsions and febrile status epilepticus

Seismic period - appearance of intractable polymorphic seizures (1-4 years)

Post-seismic period - improvement of seizures, residual mental and neurological abnormalities (2-6 years)

SCN1A mutation - alpha 1 subunit of sodium channel- 35-100%,
denovo

Familial- cases associated with mutations in SCN1A or GABRG2 demonstrate milder phenotypes. The mutant channels- show attenuated inward sodium currents



EEG - changes range initially from being normal, later develop photo paroxysmal discharges, spike/poly spike slow waves. Later abnormalities of background are noted.

Prognosis - seizure control is poor irrespective of medications used. Severe psychomotor delay/regression is part of natural history. The progression halts around 12 yrs of age *Carbamazepine, Phenytoin and lamotrigine are contraindicated*

Infantile spasms

Infantile spasms are clusters of sudden, brief bilateral tonic contractions of the axial and limb muscles in clusters.

Infantile Seizures- Review

1841- West described this epileptic encephalopathy. The epileptic nature was described by 1950. The term Hypsarrhythmia was coined in 1952 to indicate chaotic high amplitude arrhythmic and asynchronous electrical activity.

Age- 3-12 months –peak 5 months with incidence of 2-3/10,000 live births

The spasms are Sudden, brief 0.2-2sec, slower than myoclonic jerks. They are flexor, flexor extensor, or extensor spasms. 1-30% have lateralizing features, have

1-30 cluster/day with 20-150 attacks per cluster. They occur on arousal, less frequently during sleep. Regression of milestones after onset of spasms is noted

Prenatal disorders	Perinatal disorders	Postnatal disorders
Neurocutaneous disorders, Chromosomal disorders - Trisomy 21, Miller Dieker Syndrome Malformations of cortical development, Congenital infections	HIE, Infections, trauma, and intracranial hemorrhage	Pyridoxine dependency, Nonketotic hyperglycinemia, MSUD, PKU, Mitochondrial encephalopathies, Infection, degenerative diseases, biotinidase deficiency and trauma

Eighty percent are Symptomatic in nature, Tuberous sclerosis is the cause in 7-25% of Infantile spasms. Cryptogenic in 10-15%, Idiopathic-5-10%. Familial cases are rare .

Pathophysiology-

Presence of increased excitatory activity and reduced serotonergic transmission has been documented. Altered Brain Adrenal axis, research studies have demonstrated that Corticotrophin releasing hormone may be responsible for epileptic spasm and cognitive deterioration.

EEG

Interictal EEG in 2/3 is chaotic, with high amplitude, arrhythmic activity

Asymmetrical , modified hypsarrhythmia is noted in 1/3. The EEG in Tuberous sclerosis may demonstrate minor changes

Lissencephaly, Aicardi syndrome- frequent burst suppression activity

Ictal EEG demonstrates high voltage generalized slow wave activity with episodic low amplitude fast activity and electrodecremental pattern in some.

Treatment

Steroids and Vigabatrin are the drugs of choice

In UK Infantile spasm study the response with respect of cessation of spasms was Oral steroids/ACTH 73% Vs Vigabatrin 54% IS in Tuberous sclerosis- Vigabatrin appears to be better.

The Cochrane review concludes that hormonal treatment resolves spasms faster and in more number of infants than Vigabatrin. The hormonal treatment may also improve the longterm developmental outcome.



Infantile Seizures- Review

Outcomes

Mortality is <5%, and is due to Underlying cause and side effects of medications

Sixty percent develop Other type of seizures, 1/2- motor disabilities. Two thirds have variable degree of Cognitive impairment. Autistic spectrum disorder is noted in some children. 5-10% will be normal predominantly from the idiopathic group. Initial aggressive management of infantile spasms with hormonal treatment definitely makes a long term difference in outcome as we are already experiencing this in our cohort of Infantile spasms

This review is not meant to be exhaustive, there are more conditions that we encounter during day to day practice which are not discussed here. They can be addressed in a separate issue The investigations needs to be individualized as there are many investigations available.

Suggested further reading

Infantile Spasms The Lancet Neurology, Volume 4, 11, 712-717

Intermittent clobazam therapy in febrile seizures. Indian J Pediatr 2005;72:31-33.

Febrile Seizures Review BMJ 2007;334:307-311

Idiopathic epilepsies with seizures precipitated by fever and SCN1A abnormalities Epilepsia. 2007;48:1678-85

Healthy Tips

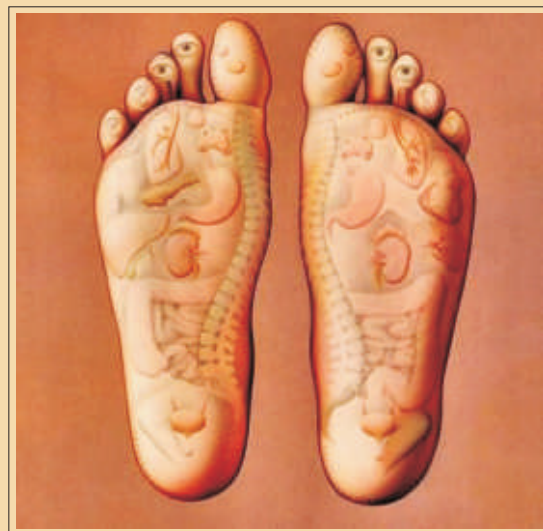
Keep Walking - health tips

Just to check this out.....

The Organs of your body have their sensory touches at the bottom of your foot, if you massage these points you will find relief from aches and pains as you can see the heart is on the left foot.

Typically they are shown as points and arrows to show which organ it connects to.

It is indeed correct since the nerves connected to these organs terminate here.



This is covered in great details in Acupressure studies or textbooks. God created our body so well that he thought of even this. He made us walk so that we will always be pressing these pressure points and thus keeping these organs activated at all times.

So, keep walking...

Obstructive Sleep Apnea

Dr Aparna Reddy,

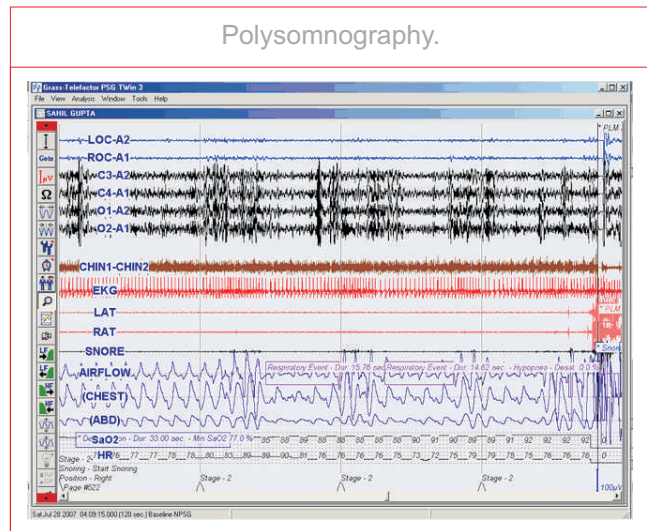
MD Paed Fellowship in Pediatric Pulmonology (Singapore)

Consultant Paediatric Pulmonologist



What is sleep study (Polysomnography)?

The sleep study or polysomnography records a variety of body parameters during sleep. It has to be conducted overnight in the hospital. The child is admitted to sleep study room. A parent is encouraged to stay with patient in the night. So as to ascertain how representative the night sleep is, compared to child's usual sleeping pattern. Moreover child will feel more assured if his/her parent is present. During admission child's medical history is taken with physical examination. The study will start when child is about to sleep. Several leads are attached including EEG, to measure brain waves and EOG – electrooculogram to measure eye and chin movement, an EKG to measure heart rate and rhythm, chest and abdominal bands to measure breathing, additional monitors to sense O₂ and C₂ levels. Monitors to record leg movements. None of devices are painful. During sleep study Apnea and Hypoapnea are recorded.



What is the treatment:

Specific therapy is tailored to the individual patient based on the severity of the OSA. Medications are generally not effective in the treatment for OSA.

Weight Reduction

Overweight persons can benefit from losing weight. Even a 10% weight reduction can significantly improve obstructive sleep apnea if obesity is the primary cause.

Surgery

Many child with OSA have enlarged tonsils and adenoids. The treatment for these children is adenotonsillectomy. Breathing assistance devices can be used in patient with severe OSA.

Many children snore during sleep. Snoring is usually harmless. However, snoring can be the first sign of a potentially serious condition known as obstructive sleep Apnea (OSA).



Obstructive Sleep Apnea

What is OSA?

OSA is a breathing disorder during sleep in which there is blockage of upper airway passage affecting normal air movement. This results in problems with sleep quality which may have widespread effects on the rest of the body.

Who gets OSA?

Children most likely to have OSA include those who have

- 1) Obesity
- 2) Central hypoventilation associated with brainstem lesions
Arnold Chari malformation
Trauma
Hemorrhage, tumor, Meningoencephalitis, hydrocephalus
- 3) Central Hypoventilation associated with neurological syndromes
Chronic pulmonary disease
OSA is reported in unexpected death
- 4) Congenital hypoventilation Syndrome

When should OSA be suspected?

Children with OSA have

- Frequent snoring
- Grunting & snorting while sleeping
- Difficulty in breathing whilst sleep followed by choking, gasping or awakening
- Restlessness during sleep with tossing & turning
- Unusual sleeping postures
- Bed wetting
- Chronic Mouth breathing
- Excessive daytime sleepiness

Why should OSA be diagnosed and treated?

OSA Can result in

- Disturbed restless sleep
- Headaches in morning
- Excessive daytime sleepiness
- Learning difficulties or poor school performance
- Developmental delay
- Behavioral or personality abnormalities
 - Poor growth including weight gain
 - High blood pressure

How is OSA diagnosed?

Diagnosis of OSA based on history and physical examination can be difficult as there are many reasons of disturbed sleep.

The current gold standard for diagnosis of OSA is the overnight polysomnography or Simply called the sleep study.

Knowledge Park

Forrest M. Bird: invented the fluid control device, respirator and the pediatric ventilator.

CPR: In the late 1950s, Dr. Peter Safar, the father of modern day CPR, pioneered the development of the ABCs (airway, breathing, circulation) of resuscitation, including "mouth-to-mouth" resuscitation. Peter Safer invented cardiopulmonary resuscitation or CPR.

In the 1960's Prof. K. Hammacher and Hewlett-Packard began development of what became the first commercially available non-invasive fetal monitor. The research took place in Boeblingen, Germany. In the spring of 1968, the first HP 8020-A fetal monitors (aptly named "The Babysitters").

In 1947, Jonas Salk at University of Pittsburgh's' Director of Virus Research, developed the Polio vaccine.

The stethoscope was invented by the French physician R.T.H. Laënnec. René Théophile Hyacinthe Laënnec is generally considered to be the father of chest medicine.

On 8 Nov, 1895, Wilhelm Conrad Röntgen discovered X-ray.

In 1944 Kary Banks Mullis, created the polymerase chain reaction (PCR).

Paediatric Oncologic Emergencies

Dr. Sirisha S,

MD Paediatrics (PGI), DNB, MRCPCH, Fellow in Paediatric Haematology Oncology & BMT (UK)

Consultant pediatric Hemato-oncologist



Approximately 60 to 70% of childhood malignancies are completely curable with current advances in diagnosis and treatment. Management of treatment related complications and supportive care are as important as specific chemotherapy.

Oncological emergencies are acute life threatening events directly or indirectly related to malignancy or its treatment that needs good anticipation and effective treatment. Some emergencies are initial manifestation as diagnosis is made, others as consequence of therapy.

They are broadly divided as follows.

Cardiothoracic emergencies:

- Superior venacaval syndrome(SVCS)/ Superior Mediastinal syndrome (SMS)
- Pleural & pericardial effusions/cardiac tamponade
- Pneumothorax/Pneumomediastinum

Abnormalities of blood counts

- Hyperleucocytosis
- Leukopenia
- Anemia, Thrombocytopenia

Metabolic:

- Tumorlysis Syndrome(TLS)
- Hypercalcemia, SIADH

Neurological emergencies:

- Spinal cord compression
- Raised intracranial pressure, altered consciousness
- Cerebrovascular accident, Seizures

Infectious

- Febrile Neutropenia
- Necrotizing enterocolitis, Perirectal abscess

Abdominal and Genitourinary:

- GI haemorrhage/GI obstruction
- Veno occlusive disease(VOD), Pancreatitis
- Haemorrhagic cystitis

Paediatric Oncologic Emergencies



In this article some of the important and relatively common oncological emergencies are discussed in brief.

SVCS/SMS:

SVCS is a rare but serious oncological emergency with an incidence of 2-8%, defined as compression of thin walled low pressure superior venacava. The term SMS is used when tracheal compression also occurs. Most common presenting symptoms and signs are cough, hoarseness, dyspnea, orthopnea, stridor, edema & venous engorgement of upper chest and face, plethora and cyanosis of face, anxiety and altered consciousness. Likely underlying conditions are non Hodgkins lymphoma(NHL), acute lymphoblastic leukemia(especially T cell ALL) and Hodgkins disease.

Chest X-Ray (Figure 1) reveals mediastinal mass. CT scan (Figure 2) or MRI demonstrates extent of tumour and compression of mediastinal structures. As positioning can be life threatening least invasive method is chosen for diagnosis. Where tissue diagnosis is not possible empirical therapy with radiotherapy/ steroids has to be instituted immediately with or without other chemotherapy agents.

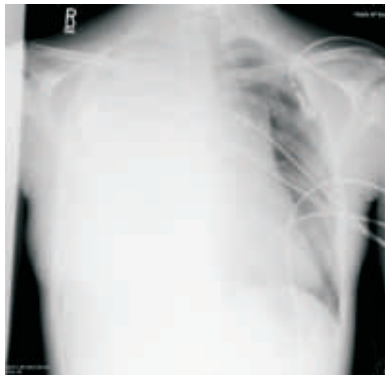


Figure 1: Chest X-Ray

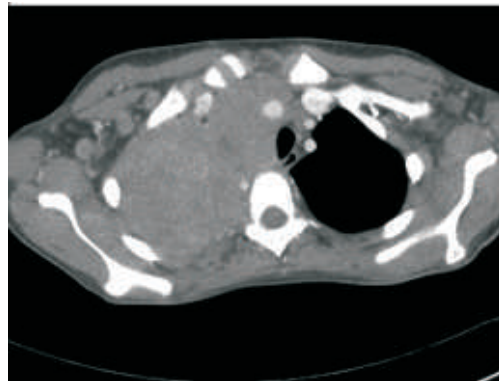


Figure 2: CT Chest demonstrating tumor in the mediastinum along with compression of trachea

Tumour lysis syndrome (TLS):

Tumour lysis syndrome is a triad of hyperuricemia, hyperkalemia and hyperphosphatemia (usually with hypocalcemia). Occurring as a result of rapid release of metabolites at rates exceeding the excretory capacity of the kidneys. TLS can occur either before starting treatment or in first few days following institution of chemotherapy. Patients with Bulky T cell or B cell lymphoma or Leukemia are at greatest risk of TLS. TLS is anticipated more in bulky disease with high serum uric acid, LDH and poor urine output.

In addition to serum electrolytes, BUN, serum levels of uric acid, calcium, creatinine, urine output, urinary specific gravity and pH should be monitored closely.

Prevention is best treatment for TLS and it requires rigorous hydration with 5% dextrose in 1/2 or 1/4 normal saline (3000ml/m²), with usage of diuretics if required to maintain urine output of 100ml/m² /hr. Allopurinol (10mg/kg/day in three divided doses for 7 days) or Rasburicase is given to control the hyperuricemia. Hypocalcemia should not be treated unless it is symptomatic. Urine alkalization is employed if required. Dialysis is indicated with renal failure or severe neurological dysfunction.



Paediatric Oncologic Emergencies

Hyperleukocytosis:

Hyperleukocytosis is defined as peripheral leukocyte count exceeding 1,00,000/mm³. It occurs in 5-20% of children with leukemias and is more often in acute lymphoblastic leukemia. In acute myeloid leukemia myeloblasts and monoblasts are more likely to cause blood hyper viscosity by virtue of their size, rigidity and stickiness. Intracerebral and pulmonary circulations are affected in hyperleukocytosis. Patient may be asymptomatic or may manifest headache, convulsions, papilledema, or intracerebral bleed in intracerebral blood hyperviscosity. When pulmonary circulation is affected they may develop dyspnea, hypoxemia or right ventricular failure. Priapism may occur in severe hyperleukocytosis.

Maintenance of hydration and aggressive management of metabolic dysfunction are vital in the management. Controlling uric acid production is essential. Red blood cell transfusion and diuretics usage should be avoided prior to cytoreduction. Cytoreduction with exchange transfusion and/or leukapheresis are indicated followed by Cytotoxic chemotherapy.

Febrile neutropenia and Neutropenic enterocolitis:

Febrile neutropenia is defined as fever of 38°C for one hour or 38.5°C of single spike with neutropenia of < 500/mm³. In addition to neutropenia, host factors like altered mucosal & skin barriers, compromised cellular and humoral immunity, nutritional deficiency are all responsible for the infections in these patients.


Key in the management of febrile neutropenia is to keep high index of suspicion and needs to be treated as medical emergency with third generation cephalosporins and aminoglycosides. Antibiotic choice may change as per the culture and sensitivity patterns of the institute and as per the focus of infection and culture results in particular patient. Antifungal therapy is indicated for prolonged febrile neutropenia treated with broad spectrum antibiotics of more than five days. Fluid resuscitation and blood component therapy is warranted where it is indicated.

Neutropenic enterocolitis is inflammation of small and large bowel. Often seen in children with malignancy during prolonged or severe neutropenic periods and is more with haematological malignancy as they develop severe neutropenia with aggressive chemotherapy. Disruption of bowel wall by chemotherapy along with neutropenia results in ulceration or full necrosis of bowel wall, presenting with abdominal pain, distension, reduced bowel sounds, vomiting and fever. Imaging of the abdomen shows thickened bowel wall especially caecum and ascending colon.

High index of suspicion is required to recognise this, as prognosis is good with prompt and aggressive management which includes bowel wall rest with nasogastric aspiration and IV fluids; broad spectrum IV antibiotics to cover gram negative bacteria and blood component therapy. Surgical intervention is indicated in case of perforation or severe gastrointestinal bleeding or uncontrolled sepsis.

Spinal Cord compression:

Spinal cord compression occurs in 2-5% of children with cancer and mostly secondary to tumours like Ewings sarcoma/neuroblastoma/lymphoma/leukemia. It presents with back pain, weakness, limping, sensory disturbance, bowel or bladder disturbances. This requires evaluation with MRI and is given immediate attention and should be started on dexamethasone 1-2 mg/kg. Some also require local radiotherapy, surgical decompression and chemotherapy. Prognosis for neurological recovery depends on duration of symptoms and degree of neurodisability at diagnosis.



Paediatric Oncologic Emergencies

If not anticipated, recognized early and treated appropriately oncologic emergencies can result in significant morbidity and death. Hence high index of suspicion and institution of immediate supportive care and early referral are essential factors for good outcome in these patients.

Suggested Further Readings

- 1) Pizzo & Poplack: Principles and Practice of Paediatric Oncology. 5th Ed, 2006
- 2) ONCOLOGIC EMERGENCIES. Pediatric Clinics of North America. 44; 4:809-830
- 3) Radiation Oncology Emergencies. Hematology/Oncology Clinics of North America. 20, 2;505-522



MRI spine demonstrating obliteration of CSF space in lumbar region with root compression

Stress - Break!..



IN THE WAITING ROOM

I was sitting in the waiting room of the hospital after my wife had gone into labour and the nurse walked out and said to the man sitting next to me, "Congratulations sir, you're the new father of twins!" The man replied, "How about that, I work for the Doublemint Chewing Gum Company." The man then followed the woman to his wife's room.

About an hour later, the same nurse entered the waiting room and announced that Mr. Smith's wife has just had triplets. Mr. Smith stood up and said, "Well, how do ya like that, I work for the 3M Company."

The gentleman that was sitting next to me then got up and started to leave. When I asked him why he was leaving, he remarked, "I think I need a breath of fresh air." The man continued, "I work for 7-UP."



Case of the month :

Unusual treatment of a movement disorder

Dr. Lokesh Lingappa, Consultant Pediatric Neurologist

Dr. Lata Bhandra, Senior Registrar

Dr. Ramesh Kancherla, Consultant Pediatric Gastroenterologist and Hepatologist

Dr. Narendra Kumar, Consultant Pediatric Surgeon

A five and half yrs old boy, first presented to our hospital with history of increasing pallor and puffiness around the eyes and face for 20days prior to admission in early part of 2007. History of breathlessness while playing noted for the same duration. Two months prior to this he had received a blood transfusion when hemoglobin had dropped to 5 gm%. He was investigated during that admission Ferritin and iron levels were low , TIBC was normal, Hb electrophoresis was normal with elevated retic count of 7%.

In the background he had history of recurrent vomiting since the age of 2 years along with failure to thrive. His weight was 18kg at 3 years and 12Kg at five and half years. He had recurrent UTIs for which was investigated with USG KUB which was normal, DMSA scan was normal. On further evaluation was found to have balanoposthitis for which he underwent circumcision at 5 years. Twice had febrile seizures, initially at 2and half years and later at 3 years, since then no recurrence.

During the admission at five and half years he was further investigated for the cause of anemia. The source of blood loss was found on Upper GI contrast study which demonstrated – Gastro esophageal reflux (GER) up to mid esophagus , the Upper GI Endoscopy showed Grade IV Oesophagitis and the biopsy documenting severe ulcerative oesophagitis, H Pylori was Negative, mild chronic antral gastritis was noted Medical management for GER was commenced with antacid(sucralfate), antiemetic (Domperidone and CiZapride). After initial improvement in anemia and weight loss once again represented with anemia few months later. At this stage parents were counselled for fundoplication. The parents were not keen for any surgical intervention.

Throughout this period since age of four and half years he also had an interesting movement disorder involving his neck. This was in the form of involuntary non rhythmic, intermittent head nodding predominantly side to side movements were noted. These were present during awake period, disappearing during sleep. He preferred to lie down to reduce these movements.

Neurologic examination was normal except for presence of these intermittent involuntary rotatory lateral movements of neck. Child was able to transiently control the movements by supporting his neck. This was diagnosed as rotatocollis. He was reviewed by multiple neurologists and Investigated while managing other problems.

The investigations included MRI of cervical spine and Brain which were normal, Nerve conduction study was normal , KF ring was negative, had normal LFT, copper and Caeruloplasmin levels. Peripheral smear for acanthocytes were negative. Trial of BOTOX was without any benefit

Born to G2 mother at term by LSCS (non progress of labour), baby cried immediately after birth. Birth weight was 3.1kgs. His developmental milestone were normal.



Case of the month :

Unusual treatment of a movement disorder

He was re evaluated in later part of 2007 as the anemia and weight loss recurred and the repeat endoscopy done this time again demonstrated grade IV Reflux with esophagitis.

This time parents were ready to go ahead with fundoplication, Laparoscopic Nissen Partial Fundoplication with gastrostomy was done in December 2007. Pre Gastrostomy the weight was 15 KG, Started on gastrostomy feed, oral intake improved, gastrostomy tube was removed, at present he is accepting oral feeds well, weight is 17.5Kgs. The movement disorder improved only after the fundoplication with near disappearance of the abnormal movements.

He also had history of recurrent urinary tract infection, 3 episodes of documented UTI, MCUG done at 5 yrs of age, suggestive of thin leucant line in posterior urethra. Cystoscopy and Circumcision: operated on 05.01.07. Operative findings: Normal, posterior and anterior urethra n DMSA scan was normal

Review of Literature

The failure to thrive, recurrent aspiration pneumonia, anemia are well reported complications of severe GER. Movement disorders reported with GER include well established Sandifer's syndrome presenting usually earlier in infancy but median age ranging from 18 to 36 month. Usually in later age group it is usually seen in children with neurologic disability.

The characteristic movement in Sandifer's syndrome is intermittent hyperextension of head often associated with rotation or tilting that may involve trunk as well. In the index patient the movement disorder was restricted only to lateral rotation movement of the neck without any hyperextension This and later age of onset made us to investigate for alternative causes for the movement disorder. The response to fundoplication suggests the rotatocollis was secondary to the severe GER the child had.

Fig 1-The Upper GI contrast study demonstrates grade IV GER



Rehabilitation services at Rainbow Children's hospital and Perinatal Centre

Occupational therapy

Physiotherapy

Speech therapy

Pediatric Occupational Therapist provides evaluation and treatments of

Gross and fine motor development.
 Feeding difficulties - oral motor difficulties and oral aversion
 Sensory integration
 Visual skills including visuo-motor coordination and
 Visuo-perceptual skills
 Self help skills such a feeding, dressing and bathing
 School skills such a handwriting, Praxis and problem solving
 Social behavioral problems

Following patients are referred to occupational Therapy

Developmental Delay
 Cerebral delay
 Dowd's syndrome
 Autism
 ADD/ADHD
 Learning disability
 Behavior disorder
 LMN dysfunction

Checklist for Occupational therapy referral

Gross Motor

Seems weaker than other children his/her age
 Does not have the endurance other children his/her age have for an activity
 Difficulty with hopping, jumping, skipping, or running as compared with others his/her age
 Appears stiff and awkward in his/her movements
 Clumsy, does not appear to know how body works, bumps into others or objects, never quite sits in char correctly
 Does not seem to understand concepts such as right, left, front, or back as it relates to his/her body
 Shies away from playground equipment. May only play on one particular item
 Poor posture (always seems to be leaning against something, shoulders slump forward)

Fine Motor

Difficulty with drawing, coloring, tracing
 Avoids fine motor activities
 Problem holding pencil, grasp may be very loose or very tight
 Printing is too dark, too light, too large, too small
 Does not seem to have a dominant hand



Rehabilitation services at Rainbow Children's hospital and Perinatal Centre

Academic

Distractible

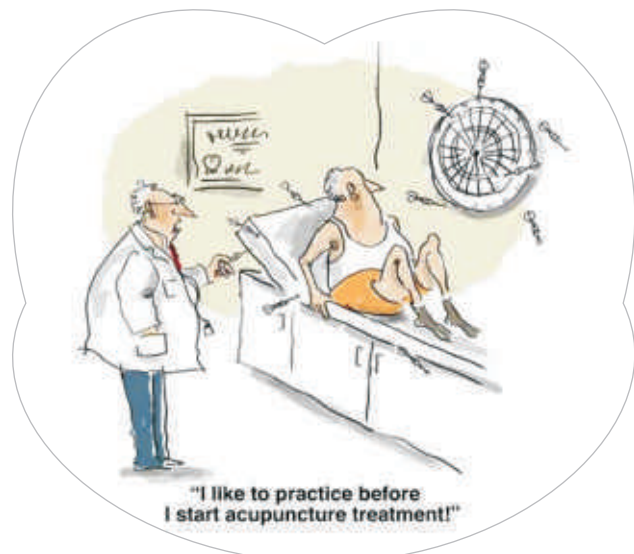
- Restless
- Slow worker
- Disorganized, messy desk
- Short attention span
- Hyperactive
- Can't follow directions
- Never completes assignments

Visual Perception

- Trouble discriminating shapes, letters, or numbers
- Can not complete puzzles appropriate for age
- Difficulty copying designs, letters or numbers
- Difficulty tracking (i.e., as in reading in a book or following teacher's arm movements)

Emotional

- Does not care to have routine changed
- Is easily frustrated
- Can not get along with others
- Accident prone
- Deals better with a small group situation or one-to-one
- Frequently involves self in other people's activities





Pediatric Neuro-Developmental Clinic Rainbow Children's Hospital

Infant spends time in Neonatal I.C.U because of problem that call for special care. While most of these problems are not long term, we know now that some children who require intensive care at birth are at a greater risk than normal for Developmental delay. However these delays can be reduced if they are identified and addressed early on.

Who are at risk for developmental delay?

Neonates with

- Low birth wt
- Premature and ventilated
- Birth asphyxia
- Congenital anomalies
- Chromosomal anomalies
- Respiratory distress syndrome
- Septicemia (infection)
- Hearing and vision difficulty
- Brain hemorrhage
- Seizures
- Nutritional deprivation

"The high risk infant follow up program" provides children who have been in NICU with periodic developmental assessments. We would like to offer this service to your family during your child's early years. We feel that both you & your child can benefit from the program. A delay/ deviation in development may be detected early, evaluated and therapy can be started if it is appropriate. This reduces the chances of further developmental delay.

"The high risk infant follow up program" will provide periodic evaluation of your child at Rainbow Children's Hospital. This evaluation will take place when your child is (6 weeks, 4 month, 9 month, 18 month and 2½ years and 3&1/2yrs).

The evaluation will include developmental screening test & a routine physical exam to see how your child is progressing in gross motor skills such as rolling, sitting, walking, fine motor such as tracking, reaching & grasping objects, eye hand co-ordination skills, language skills such as cooing, babbling, first word, social interaction such as smiling & reacting to strangers.

Thus, 'high risk infant follow up program' provides comprehensive evaluation of follow up services for children from birth to age 3½ years. Our services are geared to improving or baby's growth development of nutritional states.

Our main objective of high risk infant follow up program is to detect any medical, neurological or developmental abnormalities & provide early intervention if needed.

High Risk Infant Follow-Up program



Dr. Lokesh Lingappa

MD., DM, MRCPCH, Fellow in Pediatric Neurology (U.K),
Consultant Pediatric Neurologist

Dr.Satish Ghanta

MD (PAED) Neonatologist (Australia)
Consultant Pediatric Intensivist (Australia)
Accredited-Bayley's developmental assessment

Dr. Pranita

MOTh
Occupational therapist

Dr.Neha

MPT
Physiotherapist

Dr. Farhana / Dr.Ravi Kumar

Speech and Language therapist

Dr. Srilakshmi

MD Psychiatry
Child Psychiatrist

Dr Brindavani

Nutritionist

Dr. Neha Gaurav, BPT, MPT, Senior Physiotherapist

She did her Bachelor's and Masters in Physiotherapy from Manipal. She was awarded the best outgoing student in her specialization. After working in varied disciplines, her interests and research work in the field of women's health and pediatrics has brought her to Rainbow hospital. Working with Rainbow for more than a year now, she specializes in taking care of children with special needs, children with or without ventilator support having cardio-respiratory problems and orthopedic issues

Dr Pranita Pilkhane, BOT, MOT, Paediatric Occupational Therapist

did her bachelor of Occupational therapy at Nagpur and master from KEM, Mumbai. She has worked as Assistant Professor at SRM College, Chennai. Her expertise includes guiding children with various neurological disabilities, Autistic spectrum disorder, ADHD, Dyslexia. She has extensive experience in managing children of various age groups and is a very dedicated Professional.

Day : Tuesday

Time : 11:00am to 2:00pm

Place : Rainbow Children's Hospital
Road No-10, Banjara Hills
Hyderabad.

For appointments Call

2331 9191, 2331 9061 to 64





Care inspires and gently reassures us. Lending us a feeling of security and support, it reinforces our connection with others. Not only is it one of the best things we can do for our health, but it feels good -whether we're giving or receiving it.

- Howard Martin



Hyderabad



Secunderabad



Vijayawada



Hyderabad : Plot # 22, Road No. 10, Banjara Hills, Hyderabad. Ph : 040 2331 9191, 2331 9061 to 64. Fax : 2339 7476.

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