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UPSURGE IN APPLICATION TO SALVAGE LIVER DISORDERS IN INDIA

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- Neonatal cholestasis (NC) constitutes 30% of hepatobiliary disorders in India. First step in approach is to differentiate between neonatal hepatitis, paucity of intrahepatic bile duct and biliary atresia.
- One in 3 causes of NC is biliary atresia.
- Biliary atresia is best diagnosed by liver biopsy and so is paucity of bile ducts.
- Excretory HIDA rules out biliary atresia, but non-excretory HIDA does not mean that it is only biliary atresia. Metabolic liver disease forms a major component of hepatocellular causes of NC.
- Early referral is the key as success of surgery in biliary atresia is best at <60 days in an expert hand.
- Newer conditions like progressive familial intrahepatic cholestasis (PFIC) are more aggressively diagnosed now.
- A previous Kasai portoenterostomy (Kasai-PE) increases post-liver transplantation (LT) surgical complications such as sepsis, bowel perforations and re-exploration though there may be no significant difference in survival.
- Surgery in PFIC - Partial biliary diversion for non-cirrhotic patients; ameliorates pruritus, improves LFT and histology; liver transplantation is indicated for end stage liver disease.
- PFIC in Indian children - PFIC accounts for 8% neonatal cholestasis and 34% of cholestasis in older children with PFIC 2 being the commonest subtype. Medical therapy is successful in majority of patients. Partial internal BD should be offered to noncirrhotic low gamma-glutamyl transferase PFIC with intractable pruritus.
- Rare etiologies for liver failure in children are being diagnosed in India, such as mitochondrial DNA depletion syndrome.
- First swap donor liver transplantation in India was done in June 2009. We have achieved a lot in viral hepatitis management - Hepatitis B vaccine coverage has improved in India; we have conquered hepatitis C cure with drugs like ledipasvir, sofosbuvir and ribavirin.
- Future directions - Spread of knowledge Pan India; increased awareness for early diagnosis of treatable disorders (Galactosemia, Wilson's disease); strengthen our molecular and metabolic research for etiology and targeted management.

MYOCLONUS - PRACTICAL APPROACH "ALL MYOCLONUS ARE NOT EPILEPTIC"

Dr PAM Kunju, Trivandrum

- Myoclonus is a sudden, shock-like contraction of a muscle or group of muscles.
- Myoclonus can be divided into cortical, subcortical, spinal or peripheral, based on the presumed source of its generation.
- Based on etiology, myoclonus may be classified as epileptic or nonepileptic (physiological, essential, or psychogenic).
- It is caused by abrupt muscle contraction, in the case of positive myoclonus, or by sudden cessation of ongoing muscular activity, in the case of negative myoclonus (NM). NM results from toxic-metabolic causes.
- In a given patient, more than one form of myoclonus may occur. For instance, in posthypoxic myoclonus (Lance-Adams syndrome), cortical myoclonus may coexist with brainstem myoclonus.
- Cortical myoclonus mainly affects the distal upper limbs and face. It can be stimulus sensitive, typically to touch. If prolonged, it is called as epilepsia partialis continua.
- Subcortical myoclonus originates between the cortex and the spinal cord and is divided into nonsegmental and segmental types. Nonsegmental subcortical myoclonus - Startle/hyperekplexia and reticular reflex myoclonus.
- Brainstem myoclonus is manifested by generalized jerks and its most striking clinical feature is

sensitivity to auditory stimuli. Two main types are (i) startle response, which may be physiologic or pathologic (hyperekplexia), and (ii) reticular reflex myoclonus.

- Segmental subcortical myoclonus e.g., palatal myoclonus - caused by a lesion in the Guillain-Mollaret triangle - (dentate nucleus, red nucleus and inferior olivary nucleus).
- Spinal segmental myoclonus is usually symptomatic of an underlying structural lesion such as syringomyelia, myelitis, spinal cord trauma, vascular lesion or malignancy.
- Epileptic myoclonus is accompanied by generalized epileptiform discharges on EEG, but the myoclonus itself may be focal, segmental or generalized.
- Generalized myoclonus can occur in the syndromes of primary (idiopathic) generalized epilepsy (e.g., juvenile myoclonic epilepsy) or in the secondary (symptomatic) generalized epilepsies (e.g., progressive myoclonic epilepsy [PME]).
- Focal myoclonus can occur in symptomatic epilepsy, in the setting of infection, inflammation, vascular disease, trauma or tumors.
- Among PMEs, slow myoclonus is a feature of SSPE. In addition to myoclonus, PMEs have dementia and cerebellar ataxia.
- Early infantile myoclonic epilepsies can be benign depending on age (neonatal, infantile or childhood). Early infantile epileptic encephalopathies (EIEE) are characterized by myoclonus and other types of seizures and EEG feature called as suppression burst pattern.
- The treatment of myoclonus depends on the underlying disorder.
- Antiepileptic drugs such as valproate, levetiracetam and piracetam are effective in cortical myoclonus. Clonazepam may be helpful in all types of myoclonus.

LEGAL ISSUES IN QUALITY CARE AND CRITICAL CARE

Dr Sudhir Mishra, Jamshedpur

- The practice as a specialist should be backed by degree and not mere experience of working in a particular specialty.
- A doctor has a right to refuse a patient, especially if the patient does not belong to his specialty, doctor may not be available for full duration of treatment, or patient has a history of misbehavior. However, it is advisable to tactfully refer the patient to a higher center, rather than outright refusal.
- A doctor cannot refuse initial care in an emergency situation.
- It is advisable to equip the PICU setup in accordance with the skill available and not treat a patient beyond one's skill level.
- It is advisable to employ trained nurses rather than trying to train them at your own setup, especially if you are catering to the seriously ill patients.
- The age of viability for preterm babies varies depending on facilities available. The resuscitation in babies born between 24-28 weeks gestation should be carefully handled in consultation with parents.
- Delegation of duty does not absolve you completely from responsibility. You are liable for the acts of trainees working under you. While traveling out of station, it is desirable to hand over the patient formally to a qualified person and inform the same to parents.
- Practice in crosspathy and cross specialty is unacceptable.

ENURESIS IN CHILDREN

Dr C Suresh Kumar, Hyderabad

- Voiding of urine in bed after 5 years of age for more than 2 times a week for 3 months is called nocturnal enuresis.
- By 5 years of age, 90-95% are dry during the day and 80-85% are continent at night.
- Enuresis may be primary (85%) or secondary (15%).
- Family history is positive in 50% of cases.
- If one parent was enuretic, each child has a 44% risk of enuresis.
- If both parents were enuretic, each child has a 77% likelihood of enuresis.
- The best approach to treatment is to reassure parents that the condition is self-limited.
- The simplest initial measure is motivational.
- Initial management - General and motivational.
- First-line management includes alarm and desmopressin.
- Further treatment - Anticholinergics and tricyclics.
- Nearly 98% will have dry nights on their own.

LIVER TRANSPLANTATION IN INDIAN SCENARIO: CHALLENGES AND WAY FORWARD

- Liver transplantation is an established modality for acute liver failure, chronic liver failure and metabolic disorders.
- The first successful liver transplantation was performed in India in Apollo, Delhi in 1998.
- Challenges in India are: Lack of cadaveric donors; late referral; low socioeconomic status; too complex.
- Solutions: a) Lack of cadaveric donors - Cadaver donation has increased over the last 3 years; ABO incompatible transplants are being increasingly performed; SWAP transplants can offer hope to some families. b) Referral - Patients are being referred earlier allowing optimizing care before transplantation. c) Low socioeconomic status - Lower costs, fixed packages and support from charities and individual donors have enhanced access. d) Too complex - Better post-op care, standardization of protocols and excellent surgical expertise have resulted in outcomes comparable to the more established centers in the West.
- More than 850 pediatric liver transplants have been performed in India. Of these, 239 have been at Apollo. The Apollo Transplant Program has now performed 2,990 liver transplants. Sanjay, the first recipient of a liver transplant in India at Apollo, Delhi at the age of 18 months, is now leading a normal life 19 years later as a medical student.

HEALTH EVALUATION OF A NEWLY ADOPTED CHILD

Dr Avinash Bhosale, Jalgaon

- There have been 16,181 adoptions in the last 4 years. Therefore, it is important to discuss health evaluation of a newly adopted child.
- Initial evaluation may require several visits to the pediatrician. It involves comprehensive health planning encompassing medical history; developmental assessment; unclothed examination; laboratory investigations; referrals to medical, developmental, mental health and dental specialists.
- Special issues to be addressed at preadoption visit - Nutrition, lactation, available community support services.
- Complete history is rarely available.
- Components of comprehensive physical examination pertinent to adoption - General appearance; vital signs; growth parameters; skin examination (infectious diseases, rashes, infestations, congenital skin abnormalities, bruises or scars); genitalia examination (testing for STD to be performed with any suspicion of abuse or sexually active); neurological examination (developmental and neurologic abnormalities).
- Assess immunization status: If the status is known, document date and age; check for serum immunity for major antigens; if the status is unknown, reimmunize the child.
- Help families to promote strong, healthy attachments within family unit.
- Role of Indian Academy of Pediatrics (IAP) - IAP should formulate a chapter and guidelines to provide information and training. Adoption and foster care medicine is an evolving subspecialty in the field of pediatrics.

CHOOSING THE RIGHT AED - DIFFERENT SEIZURES, DIFFERENT DRUGS

Dr Sudhindra Aroor, Bengaluru

- **Before starting any AED**: Confirmation of seizure(s), classify the epilepsy (seizure type, syndrome), rule out other etiologies, appropriate AED - start low, go slow.
- **Starting of AEDs:** Start first-line drug at low dose and gradually optimize until seizures stop; if seizures persist, review the diagnosis and etiology; if there is definite seizure, then plan second monotherapy or go for rational polytherapy.
- **AED consideration:** *Numerous variables should be considered including* - AED specific variables (seizure- or epilepsy-syndrome, efficacy, adverse effects, pharmacokinetics, formulations, and so on), patient-specific variables (genetic background, sex, age, comorbidities, socioeconomic status), nation-specific variables (AED availability, AED cost); AEDs as a clinical test for confirming a diagnosis of epilepsy has no justification.

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