

RESULTS OF STAPES SURGERY

*Dr. Abhishek Sharma, Dr. Produl Hazarika, Dr. Dipak Ranjan Nayak
Dept. of ENT Kasturba Medical College, Manipal India*

Typically, otosclerosis presents as a slowly progressive conductive hearing loss in the third to fourth decade. Many well-documented studies have demonstrated excellent long-term hearing results with stapedectomy. Although stapedectomy is highly effective, the rare complications may be devastating. Despite a widespread use of stapes surgery, little is known about the long-term durability of hearing results. The present study provides data over a long time frame (5 years) on hearing changes following surgical treatment. During a 5-year period (2002-2006) stapes surgery was performed in 60 consecutive patients in our institution. Postoperative evaluation was performed in the same hospital. The minimum follow-up time at the latest visit was 6 months. At the long-term follow-up, we found that the operative result was very well maintained. Most of the patients reached postoperative (6-12 months) air-bone gap closure less than 20 dB. The result was maintained by the patients. Stapes surgery gives a better social life for the hearing-impaired patient and delays the need for hearing aid in most patients.

A CASE OF MAYER-ROKITANSKY-KUSTER-HAUSER SYNDROME

*Dr. Ranabir Pal¹, Dr. Ankur Barua²
Associate Professor¹, Assistant Professor², Dept. of Community Medicine,
Sikkim Manipal Institute of Medical Sciences, Gangtok, Sikkim.*

14-year-old adolescent, with ectomorphic innocent feminine appearance, presented with congenital absence of right thumb, scoliosis, severe anaemia, primary amenorrhea. She had normally developed breast without any other secondary sexual characteristics. She had history of recurrent episodes of abdominal pain and burning micturition with fever. There was no vaginal orifice. In the genital area there was a pair of ill developed labia majora fused at upper end with a whirling pattern containing inside the urethral opening. The lower end of genitalia had loose skin folds containing adequate amounts of rugae with a raphe mimicking the texture of scrotal skin. Diagnostic work-up included routine haematological studies, skeletal radiography, renal ultrasonography, and renal scan to demonstrate possible associations between the congenital genitourinary and skeletal anomalies. Ultrasound imaging studies confirmed that she had structures close to uterus with hematometra, but vagina and ovary were not found. Sonologically left kidney was non-visualized. Renal scan confirmed agenesis of the left kidney and hypoplastic parenchymally insufficient right kidney. Cytogenetic karyotyping revealed a 46, XX karyotyping and the presence of Barr chromatin body. Based on the clinical examination and investigations, a diagnosis of Mayer-Rokitansky-Kuster-Hauser syndrome was established with associated limb and skeletal deformities.

ATYPICAL PRESENTATION OF A CASE OF Hb E/̂⁰ THALASSAEMIA

*Dr. Ranabir Pal¹, Dr. Ankur Barua²
Associate Professor¹, Assistant Professor², Dept. of Community Medicine,
Sikkim Manipal Institute of Medical Sciences, Gangtok, Sikkim.*

A 6 year old girl of Hb E/̂⁰ Thalassemia presented with moderate anaemia, mild hepatosplenomegaly and normal phenotype without any classical feature of mega hepatosplenomegaly or bronzy discoloration of skin along with typical facial changes. This patient of Hb E/̂⁰ Thalassemia with normal phenotype had been followed up in the line of management of ̂⁰ Thalassemia. It is very important to generate awareness among our fraternity regarding subtle presentation and importance of early detection of E/̂⁰ Thalassemia in the cases of unexplained anaemia. Otherwise, they develop irreversible organ damage due to iron overload by iron supplementation. Earliest diagnosis followed by routine follow up as well and timely intervention can help in better prognosis in this treatable disease. Moreover, health education at all levels to care givers and peers would help these patients to lead a productive life in the line of "live with the disease" phenomenon. Professionals related to Community Pediatrics, Clinical Epidemiology and Community Genetics, functioning in unison, has a dominant role to play in preventing the fatal outcome of this genetic disorder.

CAPILLARY HAEMANGIOMA OF NASAL CAVITY

*Dr. Rakesh V., Dr. Dipak Ranjan Nayak, Dr. Balakrishnan, Dr. Kailesh Pujary & Dr. Rohit Singh
Dept. of ENT, Kasturba Medical College, Manipal.*

Although the head and neck is not an uncommon region, the nasal cavity is an extremely rare site for capillary haemangiomas and only few cases have been earlier reported in literature. We report a case of 38 year old female who presented to us with epistaxis and nasal obstruction and was managed successfully with KTP-532 laser assisted excision of the haemangioma. We feel that although capillary haemangioma of nasal cavity is extremely rare, it should be considered as differential diagnosis of bleeding lesions of Nasal cavity.

DEXTROCARDIA WITH SINGLE VENTRICLE

*Dr. Ranabir Pal¹, Dr. Ankur Barua²
Associate Professor, 2 Assistant Professor, Community Medicine,
Sikkim-Manipal Institute of Medical Sciences. 5th Mile, Tadong, gangtok, sikkim, india. 737102*

A 16 years old short stature mesomorphic girl presented with cyanosis, clubbing on twenty nails, progressively increasing generalized weakness, fatigue, breathlessness without exertion. On palpation, right para-sternal lift, systolic thrill and bruit was noted over pre-cordium. On auscultation 1st heart sound was normal; 2nd heart sound was single and loud. The ejection systolic murmur grade III was found in 'Pulmonary area'. Pulse oxymetry showed 67 % Oxygen saturation. General systemic examination delineated no other incidental abnormal or ambiguous visceral arrangement or organomegaly. Haemoglobin level was 10.2 gm Gm/dL with few reticulocytes and nucleated red cells and microcytes. On chest radiography, a bulge on upper right border of cardiac silhouette in the postero-anterior projection suggested a rudi-

mentary outflow chamber. Pulmonary vasculature was oligoemic suggesting pulmonary outflow obstruction. Electrocardiogram findings were non-specific and did not match with any prototype. There was left axis deviation, normal PR interval and no gross changes in QRS complex in limb leads. P waves were spiked in VR, VF, V₁, V₂. Normal in I, VL, Bifid in II, III, VF, V₃₋₆ - this is a characteristic finding in univentricular heart. A negative P wave in lead I indicated reversed atrial arrangement confirming dextrocardia (atrial situs inversus) QRS complexes were characteristically larger in right chest leads and progressively smaller from V₄₋₆ in left sided leads. In all chest leads there were also additional findings of predominantly rS pattern in QRS complex. There was 'W' pattern of QRS complex in Lead II and V₂. T waves were deeper in I, VR, VL, all precordial leads, taller than normal in others. Absence of ventricular septum was the principal echocardiographic sign with morphology of a normal left ventricle and two inlet valves and the right ventricle was represented by a small rudimentary outflow chamber. It appeared that aorta was arising from the rudimentary chamber and stenosed pulmonary artery from the main chamber. The findings clinically lead to a diagnosis of dextrocardia with single ventricle.

PARTIAL DENYS DRASH SYNDROME, A RARE DISORDER WITH A RARE ASSOCIATION OF CONGENITAL CYTOMEGALOVIRAL INFECTION WITH CHORIORETINITIS

*Dr. Jayashree K, Dr. Suneel C Mundkur, Dr. Sri Kiran Hebbar
Department of Pediatrics, K.M.C. Manipal, India.*

Congenital nephrotic syndrome is an uncommon cause for nephrotic syndrome in children. Apart from Finnish type, intrauterine infections are other important etiological factors. Denys - Drash syndrome is a rare entity characterised by triad of congenital nephropathy, Wilms tumor and intersex disorders. Incomplete forms are known to exist where congenital nephropathy exists either with Wilms tumor or with intersex disorder. Children with 46XX karyotype can have no intersex disorder. As Wilms tumor develops only later in life, this type of Denys - Drash syndrome may not be considered as a cause in a case of congenital nephropathy initially. Hence all cases of congenital nephropathy without intersex disorder should be considered for molecular study, for detecting WT1 gene mutation at 11p13 locus to diagnose in advance. Bilateral nephrectomy with renal transplant can be life saving in these children. We report a case of incomplete form of Denys - Drash syndrome with no intersex disorder who also had evidence of CMV infection with associated retinitis, which also can cause congenital nephrotic syndrome. To best of our knowledge we have not come across any similar case in the literature.

"UNUSUAL PRESENTATION OF SPINAL CORD INFARCTION IN A PATIENT OF SICKLE CELL ANEMIA : A CASE REPORT"

*Dr. Bharti Ganvir, Lecturer, Deptt. Medicine, Dr. Sindu, Resident, Deptt. Medicine,
Dr. Namita Jajoo, Resident, Deptt. Medicine, Dr. A.P. Jain, Professor & Hod., Deptt. Medicine
Vascular disease of the spinal cord occurs with less frequency than in the brain. Its manifestations are similar, however, and are often abrupt in onset, dramatic in scope, and frequently disabling. The most common serious neurological complication of sickle cell anemia is occlusive vascular disease with central nervous system infarction. The parenchymal lesions are most often located in the brain, chiefly within major cerebral arterial boundary zones. Spinal cord infarction is extremely rare. Here we report a patient with sickle cell anemia who developed an ischaemic myelopathy secondary to sickle cell anemia.*

MANIFESTATION OF DHAT SYNDROME IN THE CONTEXT OF PAKISTAN

*Nashi Khan, University of Health Sciences, Lahore, Pakistan
Prof. Dr. Ruksana Kausar, Fatima Jinnah Women University, Rawalpindi, Pakistan
Prof. Dr. Haroon Rasheed Choudhry, P.I.M.H., Lahore, Pakistan.*

Dhat Syndrome also called semen loss syndrome, is a culture bound syndrome most commonly prevalent in Indian sub continent. This was a pioneering study on a tabooed area in Pakistan. Main objective of the study was to examine manifestation of the syndrome in the Pakistani context. 318 Dhat Syndrome patients comprised the sample and they were recruited from private practice of Hakims, Homeopaths, Fertility Specialists and General Practitioners comprised the sample. Especially designed interview schedule and Dhat Syndrome Symptom Checklist (DSSC) were used to gather data. The patients were interviewed at the clinics of professionals. Analyses revealed that patients were consulting both medical and traditional healers. Majority were single, young, less educated with poor socioeconomic status. They reported masturbation and exposure to pornographic material the major causes of Dhat Syndrome. Though the patients reported physical, psychological and sexual symptoms, manifestation in majority was in physical form. Sex being a neglected and tabooed area in Pakistan, this is the ever first systematic empirical study on the subject. The findings from the study have very important implications for men's sexual health and medical professionals.

INCIDENCE & PATTERN OF NASH IN INDIAN AIRLINES

Dr. P.K. Shrivastava, M.D (Medicines), FIMSA, Indian Airlines Ltd, New Delhi
Non-Alcoholic Steatohepatitis (Fatty Liver) is an important public health problem because it is more preventable than Hepatitis B, C & Alcoholic liver disease. It is not a harmless condition and may cause fibrosis and cirrhosis. Global prevalence of disease is 10-39% of normal urban population. Usually this condition is found in insulin resistant and obese but many times in normal individual also, and is diagnosed by chance. Present study is aimed to know NASH IN Indian Airlines staff who are rich, well nourished and sedentary workers. Raised SGOT, SGPT mild hyperbilirubinemia and GGT were common findings. Incidence of fatty liver was 22% and NASH was 16%. Which are significant. There is increased intra hepatic production of FFA from glucose which is not taken up by peripheral adipocytes and myocytes. Metabolites of excess lipid stores in liver cell triggers inflammatory response leading to fibrosis. Rosiglitazone, Metformin, Gemfibrozil, Ursodeoxy cholic Acid, Vitamin E along with weight loss form the principles of treatment. The results remains variable.