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Sri Ramakrishna
Hospital (Multi-Speciality)

pulse

Happenings at Sri Ramakrishna...

49 Years of Excellence

DONATE ORGAN SAVE LIVES
AUGUST 13th

WORLD ORGAN DONATION DAY

Shri.D.Lakshminarayanaswamy
Managing Trustee

I am filled with immense pride and gratitude for the growth and innovation we have achieved together. I am delighted to be a part of the team that has made remarkable progress and achievements over the years. Our commitment in providing exceptional healthcare and serving our community has always been stronger, and I am proud of the strides we have taken together.

We recognize the profound impact that organ donation can have on the lives of individuals and their families. On this **World Organ Donation day,** we celebrate the selfless donors who gave the gift of life and renew our commitment in promoting awareness and education about organ donation.

I encourage everyone to consider becoming an organ donor. By registering as an organ donor, we have the power to give the gift of life and make significant difference in the lives of patients and their families. Together, we can make a difference and give the gift of life to those in need.

Dr. S. Rajagopal
Medical Director

Sri Ramakrishna Hospital has always been a forerunner in conducting diverse academic programs alongside its clinical achievements. The emphasis on clinical club meetings, where we discuss interesting cases adds an enriching dimension to the professional development of the team.

This month's Special focus on **Surgical Gastroenterology and Obstetrics & Gynaecology** demonstrates a strong commitment to staying updated with medical advancements and addressing a broad spectrum of healthcare needs which in turn benefits the medical professionals and also enhances the overall quality of patient care.

We are committed to promote the health and well-being of mothers and babies, and breast feeding plays a vital role in this mission. This **World Breast feeding Week** provides us with an excellent opportunity to highlight the importance of breast feeding and also by supporting our community in understanding the benefits involved.

Organ donation is an extraordinary act of compassion that can save and transform lives. One donor can save up to eight lives and enhance the quality of life for many more through tissue donation. Despite remarkable advancements in medical science, the demand for organ transplants continues to outpace the supply. On this **World Organ Donation day,** it is imperative that we raise awareness and encourage more people to donate organs and make a lasting impact on those in need.

Introducing the Future of Healthcare: All-in-One Healthcare Kiosk

We, Sri Ramakrishna Hospital, excited to announce that we have launched All-in-One Healthcare Kiosk, a ground breaking innovation in healthcare technology and first of its kind in Coimbatore. This comprehensive platform integrates various healthcare services into a single, user-friendly terminal, revolutionizing healthcare solutions for both healthcare providers and patients.

Comprehensive Healthcare Solution

Our All-in-One Healthcare Self-Service Kiosk streamlines numerous processes, including:

- Registration with online consents
- Capturing patient photographs
- Token and patient barcode printing
- Billing process
- Printing of OP cards
- UPI payments
- Debit/credit card payments
- Advance Payments
- Appointment scheduling
- Patient feedback, and many more

Benefits

The All-in-One Healthcare Kiosk offers numerous benefits, including:

- Timesaving and user friendliness
- Accurate and reliable access
- Enhanced patient engagement
- Improved health outcomes
- Reduced healthcare costs
- Increased accessibility
- Streamlined patient crowd
- Improved patient satisfaction
- Convenient billing and payment options
- Enhanced patient-provider communication
- Enhanced data security and privacy
- Real-time insights and analytics
- Integration with existing healthcare systems
- Scalable and flexible solution

Impact

In a period of last six months, our multiple All-in-One Kiosks have made a positive impact on many people, with over 17,000+ unique patients using it happily. We are proud to be at the forefront of healthcare innovation, and we are looking forward for constantly improving healthcare outcomes with latest innovations and technologies.

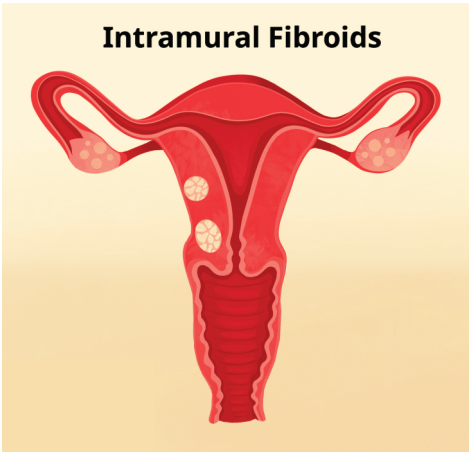
Multiple IVF Failures

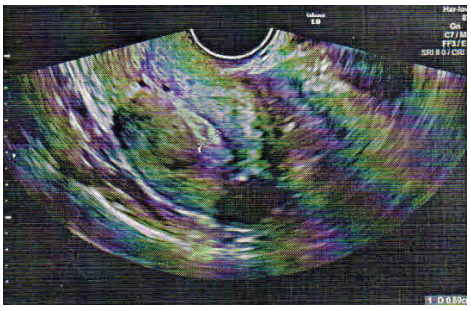
Case Report: A 39 year old female, married for 8 years consulted us on 13.09.2022.She was referred from an IVF center, with H/o repeated IVF failures for further management.

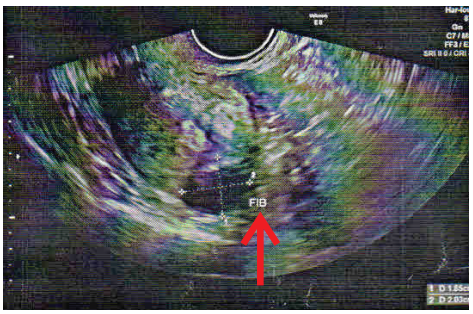
She underwent Laprascopic and Hysteroscopic myomectomy in 2017 & 2019 respectively, following which she had undergone five cycles of oocyte retrieval and 8 cycles of embryo transfer which resulted in 2 biochemical pregnancies. She was on insulin for TYPE-2 DM. chromosomal karyotyping of both partners were normal. After third ET failure ERA was performed to time embryo transfer. Husband was normozoospermic.

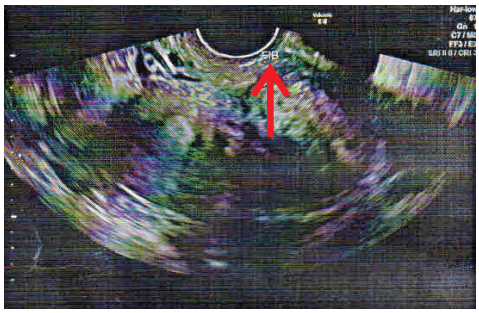
Here we had to deal with a case of multiple IVF failures, with increased age of both partners.

On evaluation, her hormonal assays were normal.









USG revealed recurrence of fibroids-two intamural fibroids were close to the cavity & multiple subserosal fibroids. SHG showed minimal cavity distension. She needed repeat myomectomy. In view of increased maternal age, she was advised embryo freezing prior to myomectomy. She underwent ICSI-program in NOV 2022. 9 ocytes retrieved, 7-embryos obtained were frozen. Dec 8th 2022, Diagnostic Hysteroscopy and Laproscopic myomectomy performed. Intraopertively during en-nucleation of intramural fibroids, cavity inadvertently opened, same sutured.

As we had to give 6 months of waiting time, she underwent one more cycle of embryo freezing in Feb 2023. 5-oocytes retrieved & 4-embryos obtained & frozen. April 2023 endometrial PRP was performed to improve endometrial receptivity. May 2023, first cycle of frozen thawed embryo transfer was done with two Grade AA blastocysts & was negative.

We offered her PGS with the remaining embryos, but she denied and she underwent second cycle of FET in June 2023. Five embryos were thawed, two good grade blastocysts obtained. AH was done on these embryos.

ET was performed according to the ERA test report which said 180 hours from time of progesterone initiation. Usually blast transfer is performed at 120 hours.Here her endometrium was receptive at 180 hours. Her Beta-HCG was positive.

During her first trimester, she had threatened miscarriage, managed conservatively. Second trimester, she had short cervix. She underwent cervical encerclage at 14 weeks. Her glycemic control and hypothyroid was taken care by endocrinologist. She delivered an alive female baby on 26.02.2024 at 37+6 weeks.

Discussion: Repeated implantation failures is defined as 3 IVF failures following transfer of good quality embryos, where in good response to ovulation induction, good quality embryos, good endometrial development &no pathology identified. Major factor is sub-optimal endometrial receptivity which inhibits embryo implantation.

During the implantation window-there is a cross talk between the endometrium and the embryo, to allow attachment and invasion of embryo.Patients with repeated IVF failures have short implantation window, which can be identified with ERA test.

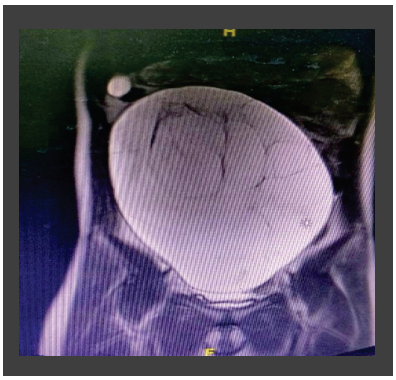
In this patient the cause of failure was intramural fibroids close the cavity and short implantation window which was identified with ERA test. RIF is seen in 10% of cases of IVF. In majority of cases no identifiable reason is found.

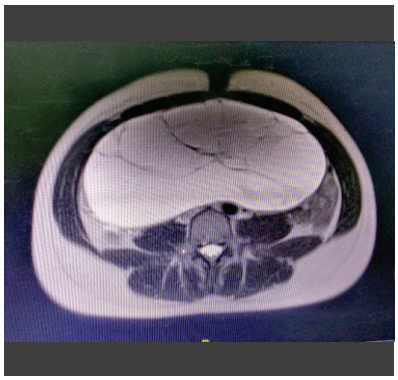
Dr.LAKSHMI.M.S
ART Consultant

How a Blooming Period Turns into Gloominess in a Teenage Girl

Case Report: A 15 year old girl attained menarche at her 13 years of age, had regular cycles for first 7 months ,presented with complaints of secondary amenorrhea for a period of 2 years , and excessive sweating associated with foul smell since 6 months and excessive hair growth over face, chest, limbs . abdominal pain since 3 months on and off which is dull type and diffuse., associated with complaints of decreased appetite, with rapid progressive abdominal distension, in a period of 20 days, having mild PCOS ,no drug intake history. The clinical examination at the time of admission revealed patient in good general condition, apyretic, with normal blood pressure and the presence of an abdominal mass approximately corresponding to 34 weeks size, extending upto xiphisternum.

On physical examination, hirsutism on her face, chest, and limbs noted. USG showed large complex right ovarian cyst with no evident free fluid, MRI abdomen plain and contrast was performed in multiple planes and images obtained Shows a large abdomino pelvic complex cyst measuring 27x 19x 8 cm (CC x ML x AP) seen displacing the uterus, postero inferiorly, the urinary bladder inferiorly, all the small bowel loops to left side and superiorly. The right ovary is not seen separately - features suggestive of large right ovarian complex cyst with minimal ascites, no obvious peritoneal or omental thickening, no paraaortic or pelvic lymphadenopathy The cyst is thin walled with multiple contrast enhancing thin internal septa and enhancing small solid regions. No blood fluid level within the cyst. No internal fat in the cyst.





A harmonal assessment showed excess androgenic activity in the form of raised testosterone levels (503 ng/ dl) raised 17OH Progesterone levels of (3.69 ng / ml). A cortisol challenge test was done showed normal serum cortisol levels (0.95 mcg / dl) ruled out Cushing syndrome. However levels of CA 125 (22.5), AFP (1.26), LDH (216), CEA (1.35), BETA HCG (< 2.39), Prolactin (12.2), CA 19.9 (3.5) were normal.

Based on these findings,a provisional diagnosis of androgen producing ovarian tumour was made with underlying non classical CAH. After discussed with the patient, the decision was to proceed to surgery.

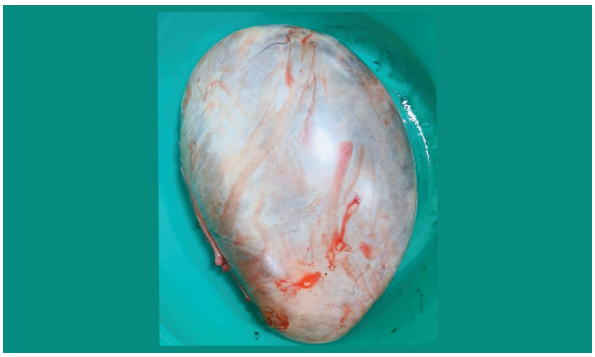
MANAGEMENT

Surgical technique and findings: The patient underwent exploratory laparotomy, with midline vertical incision extended above supraumbilically, revealing a large solido cystic right ovarian mass occupying the entire abdomen. Right fallopian tube seems stretched over the mass.

Left ovary and tube were normal: Minimal ascitic fluid seen which is blood stained, aspirsted and sent for cytology .
Pouch of douglas - free

No omental / peritoneal deposits seen, proceeded with right salphingoophorectomy and specimen retrieved enblock, and sent for Histopathology.

On gross examination: A right ovarian mass of size measuring 25x 19.5x 11 cm. External surface is vaguely bosselated. Cut surface is predominantly cystic and multilocular cyst, draining serous fluid, wall thickness is 0.2 to 0.6 cm. Focal solid area is noted, measuring 2x 2 x 1 cm . Cut surface of solid area is soft to firm and grey brown to pale white. No papillary excrescences are seen. Attached fallopian tube measuring 9 cm in length and 0.5 cm in diameter unremarkable.A 10 ml of ascitic fluid which is haemorrhagic sent for cytology, the smears prepared from the specimen is stained with Giemsa and H & E stains – shows peripheral blood elements. No malignant cells seen.

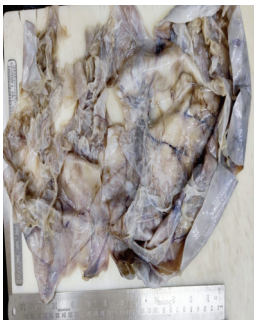


On microscopic examination: Shows ovary with a neoplasm having variable cellularity and cystic changes composed of admixture of sertoli cell components, leydig cell components and primitive stromal components. The Sertoli cell component is arranged in cords , hollow and solid tubular pattern, having scanty eosiniphilic cytoplasm , uniform nuclei, small nucleoli and powdery chromatin

Primitive stromal component is composed of round to spindle shaped/ stellate cells having scant eosinophilic cytoplasm, high N: C ratio, Pleomorphic mitotically active nuclei, condensed chromatin and inconspicuous nucleoli. Background is myxoid at places with curvillinear thin blood vessels. Leydig cell component is seen in clusters . No necrosis seen. Fallopian tube is unremarkable.

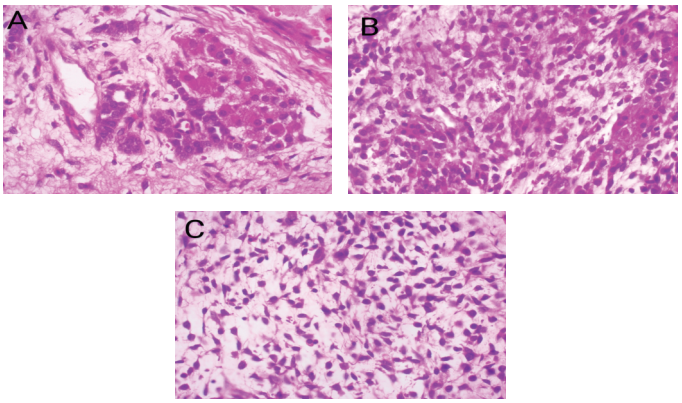
Based on gross and microscopic findings it is diagnosed as moderately differentiated Sertoli leydig cell tumour with no capsular breech

Based on clinical and staging laparotomy findings tumour classified as stage 1 C



Histopathology

A. Sertoli cells on – left side with eosinophilic cytoplasm and powdery chromatin
Leydig cells on – right side seen in clusters
B. Sertoli rich mixed cell component
C. Primitive stromal component



Outcome and follow up

Histopathology of the tumour showed grade 1 moderately differentiated Sertoli leydig cell tumour, FIGO Stage 1C, as no capsular breech and cytology negative for malignant cells. Postoperatively total testosterone levels dropped to normal range (0.642 nmol/ L), with 17OHP remaining mildly elevated (2.49 ng / ml), thus confirming the presence of underlying non classical CAH, on follow up, patient resumed her menstrual cycle, reduction in facial hair growth, reduced bromohidrosis, patient was counselled regarding the need for steroid treatment, and started, with regular follow up of the patient.

Discussion: Sertoli leydig cell tumour of the ovary form 20 % of the sex cord-stromal tumours, < 0.2% of ovarian tumour's, mostly present in younger women, they are usually unilateral, less aggressive in terms of malignant transformation, and hence optimal surgical management eradicate the tumour completely and thus helping to preserve fertility. During follow up , testosterone levels should be regularly monitored to ensure complete resolution of the tumour. The exact incidence of ovarian tumours in women with non classical CAH is not yet fully defined, it has been hypothesized that hyperandogenism in CAH is accompanied by peripheral aromatisation, leading to secondary hyperestrogenism and increased secretion of LH , stimulating the growth of ovarian interstitial tissue although the presence of PCOS in this patient seen.

Conclusion: As our patient had reduction in facial hair growth, resumed menstrual cycle post surgical, ovarian tumour was likely the major cause of virilization, although elevated 17 OHP even after surgery, concludes co existence of non classical CAH.

Dr.SUGANYA.R
MBBS, DNB (OG), DRM(Germany)

Consultant in Infertility

Amenorrhea – A Search for the cause!!

Evaluation of a young patient presenting with amenorrhea is a clinical challenge wherein one often comes across atypical clinical scenarios. The importance of an individualized and careful evaluation in arriving at the diagnosis is highlighted in the case described.

Case History: 17 years old female presented with amenorrhea since the last 10 months. She was on treatment for the same on and off without any response to therapy. There was history of severe acne and excess body hair since the last 1 yr. She had achieved menarche at 13.5 years. Her initial menstrual cycles were regular but she developed oligomenorrhea which finally progressed to amenorrhea.

She is the 3rd child born of 3rd degree consanguinity with a normal and uneventful birth and development history. There was no family history of neonatal deaths and infertility. Her height was 146 cm for a Mid Parental Height of 156 cm and weight was 42.3 kg. Short stature was confirmed on her growth chart. (FIG.1)

On examination there was no dysmorphic features. Her blood pressure was 110/70 mmHg. There was cystic acne on face and trunk. She had features of virilization like hirsutism(FerrimanGalleway score = 18),hoarse voice, temporal hair recession and muscular built. Her Tanner staging was A3 B1 (atrophy) P5. Her external genitalia examination was suggestive of clitoromegaly(3 cm) with mild posterior labial fusion. There were no palpable gonads or hyperpigmentation. Rest of the examination was normal.

The clinical diagnosis at this stage was "SECONDARY AMENORRHEA WITH VIRILIZATION".

Her laboratory investigations revealed; FSH - 6.83 mIU/L; LH - 5.15 mIU/L;Estradiol - 48.041 pg/mL;Prolactin - 12 ng/mL; Testosterone - 399 ng/dL (60-90); 17 OHP - 213.7 ng/mL (<0.9); 8 am Sr.cortisol - 28.8 mcg/dL (5-20) and normal serum electrolytes. The serum cortisol post overnight Dexamethasone suppression test (ONDST) was 1.4 mcg/dL and the high initial cortisol levels were attributed to the effect of oral contraceptive pill use.Her ultrasound was suggestive of normal sized uterus with thin endometrium; normal ovaries with multiple follicles; no abdominal mass.

This narrowed down the differential diagnosis to virilizing tumor v/s Classical congenital adrenal hyperplasia. Her CT adrenals was suggestive of Bilateral diffusely bulky hyperenhancing adrenal glands suggestive of adrenal hyperplasia without evidence of focal adrenal mass (FIG.2) thus confirming the diagnosis of Simple Virilising form of Classical Congenital Adrenal hyperplasia.

She was started on treatment with steroids (Hydrocortisone and Fludrocortisone) and the doses were titrated to achieve near normal testosterone levels. She has started menstruating again. Her acne has resolved; the clitoromegaly persists. Her genetic report confirmed the clinical diagnosis of Congenital Adrenal Hyperplasia. (Homozygous mutation in CYP21A2). (FIG.3) She needs to be on lifelong treatment with steroids.

Congenital Adrenal Hyperplasia (CAH): CAH is a family of autosomal recessive disorders resulting in deficiency of one of the several enzymes of adrenal steroidogenesis.

More than 90% of CAH cases are due to deficiency of 21 hydroxylase enzyme which is encoded by the CYP21 gene on chromosome 6. There is a broad spectrum of clinical manifestations, depending on the gene mutation. Accordingly CAH due to 21 hydroxylase deficiency presents as;

1. Classic CAH:
a. Salt wasting CAH –Severe form of the disease due to complete lack of enzyme activity. Females are born with ambiguous

genitalia; both genders are prone to salt wasting crisis in the first few weeks of life. This is now detected routinely by the newborn screening programs measuring neonatal 17 hydroxyprogesterone (17OHP) levels.

b. Simple virilizing CAH – Partial activity of the deficient enzyme prevents the occurrence of life threatening salt wasting crisis at birth. Nevertheless, females present with virilization and males present with early onset puberty.

2. Nonclassic CAH: Mild forms of enzyme deficiency present with mild to moderate hirsutism, acne, menstrual irregularities and decreased fertility in women often mistaken for the more common Polycystic Ovarian disease.

Diagnosis: In the absence of prenatal screening, genital ambiguity or a salt wasting crisis will generally alert pediatricians to most cases of severe forms of CAH. But often the other forms of CAH cases get missed till late into the disease course, when they seek treatment for virilism, hirsutism, acne, menstrual irregularity or infertility. The key diagnostic test is to measure the baseline 17 hydroxyprogesterone levels and confirm the same by doing a Synacthen stimulation test.

Management: Treatment involves careful use of steroids [glucocorticoids (GC) ±mineralocorticoids(MC)] to suppress the excess ACTH levels; thereby decreasing the androgen synthesis. Both undertreatment and overtreatment with steroids can be hazardous. Long term management needs careful clinical and laboratory monitoring.

Take Home Message:

- Various endocrine and non endocrine conditions can present with amenorrhea.
- Thorough history and clinical examination helps to narrow down the differential diagnosis.
- Investigations help to confirm/rule out the differential diagnosis.

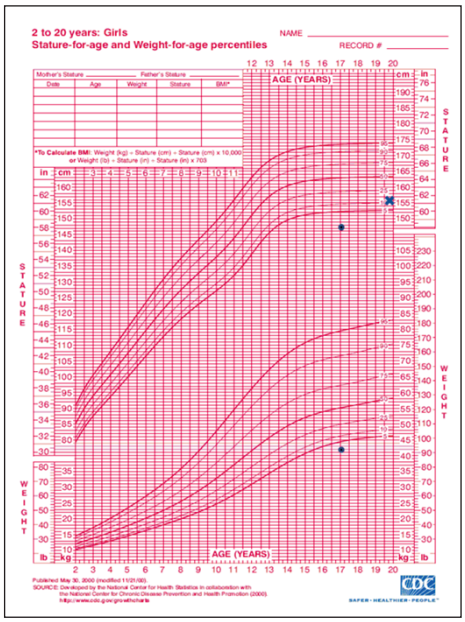


Fig: 1

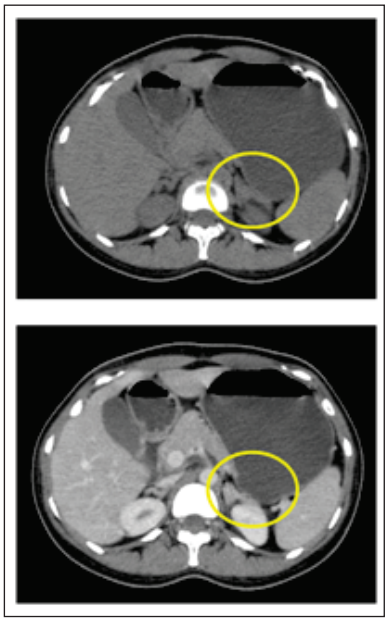


Fig: 2

Test Results and Interpretation

PATHOGENIC VARIANT CAUSATIVE OF THE REPORTED PHENOTYPE WAS DETECTED

Summary of Variants (NGS)

Gene and Transcript	Exon/Intron Number	Variant Nomenclature	Zygosity	Classification	Disease	Inheritance
CYP21A2 (NM_000500.9)	Exon 4	p.I173N c.518T>A p.Ile173Asn	Homozygous	Pathogenic	Adrenal hyperplasia, congenital, due to 21-hydroxylase deficiency	Autosomal recessive

Summary of Variants (MLPA)

The same homozygous variation c.518T>A(p.Ile173Asn) was detected within the detection limits of MLPA, in the *CYP21A2* gene of this subject.

Fig: 3

Dr.SHOBI ANANDI.V
MD, DNB, Fellowship in Paediatric Endocrinology
Consultant Pediatrician & Pediatric Endocrinologist

Paediatric Liver Transplant - The way forward

Introduction: Professor Thomas E Starzl performed first Paediatric Liver Transplant in 1960's, with first long term survival obtained in a child, transplanted in 1970 for Biliary Atresia. In 1998, India's first Paediatric liver transplant was performed in Delhi for a boy from Tamil Nadu with Biliary atresia who now survives to become a Doctor.

Paediatric liver transplantation positioned at the very birth of the Hepatic transplantation surgical speciality, which now has become the Standard of care for various ESLD in both children and adults.

Types of Transplant

Living Donor (LDLT)

- Decreased wait time
- Better survival of graft

Deceased Donor (DDLT)

- Advent of split liver transplant

Auxiliary liver transplant with implantation of a partial graft without fully removing the native liver, as suitable option for ALF and metabolic disorders without cirrhosis as it offers a chance for immunosupression free life in case of native liver regeneration. However not very popular due to the toxic and metabolic effects of the diseased liver in ALF.

Breakthrough in Liver Transplant: Paediatric liver transplant has programmed beyond ABO blood group barrier. Despite concerns about liver graft rejection, antibody mediated rejection however with the introduction of Rituximab prophylaxsis before transplant, B.cell desensitization strategies including plasmapheresis, IVIG have improved the survival rate. Children younger than 1year of age may not require these strategies as blood group isoagglutinin titres are low and complement activation system is not robust.

Size matching determined by graft to recipient weight ratio (GRWR), is a crucial determinant for graft suitability and ideal ratios of 0.8-1 are targeted. Larger for size grafts are problematic in small infants due to compromised portal venous flow and small abdominal cavity. Area of split liver grafts and delayed abdominal closure using mesh skin closure help circumvent abdominal compartment syndrome.

A 2.5years old boy our follow up Post DDLT child for Biliary atresia post failed Kasai with Liver failure underwent the First Split Liver Transplant in Coimbatore. He had acute rejection in the first post Op week, which settled with adequate immunosuppression. In 2nd post op week, child developed perforation requiring re-exploration. Gradual nutritional rehabilitation was done, he is on regular follow up with no major complications and minimal immunosupression.

INDICATIONS

Cholestatic disorders

- Extrahepatic Biliary Atresia
- Alagille Syndrome
- PFIC
- Sclerosing Cholangits
- Caroli syndrome

Acute liver failure

- Undetermined cause
- Hepatitis non ABC
- Drug toxicity like Paracetamol
- Wilson disease
- Autoimmune hepatitis

Liver tumour

- Hepatoblastoma
- Hepatocellular carcinoma
- Infantile Hemangio-Endothelioma

Others

- Budd chari syndrome
- Cryptogenic liver cirrhosis
- Infantile copper overload

Metabolic liver disease

With cirrhosis

- Antitryspin deficiency
- Wilson disease
- Tyrosinemia
- Neonatal hemochromatosis
- Cystic fibrosis
- Glycogenosis type IV
- Primary bile acid synthesis defects

Without cirrhosis

- Hyperoxaluria
- Crigler- Najjar syndrome
- Urea cycle defect
- Familial hypercholesterolemia type II A
- Glyogenosis type IA
- Hemophilia A&B
- Protein C deficiency

CONTRAINDICATIONS

- Hepatocellular carcinoma with Extrahepatic disease & Rapid progression
- Generalized extrahepatic malignancy
- Uncontrolled systemic infection
- Severe multisystemic mitochondrial disease
- Nicmann - pick disease type C
- Severe porto - pulmonary hypertension not amenable to treatment

Our Liver Transplant team with our first Paediatric Transplant Patient

Dr.INDHIRADEVI.B
MBBS, MD (Pediatrics), IDPCCM (PICU)

Consultant Paediatric Intensive Care (PICU)

Dr.KRISHNA SAMEERA.G
MD (Pediatrics), IDPCCM (Pediatric Critical Care)

Consultant Paediatric Intensive Care (PICU)

Sri Ramakrishna Hospital with Health Basix launched

Early Intervention & Child Development Center at

Sri Ramakrishna Central School

Sri Ramakrishna Hospital, in association with, Health Basix, a pioneer in child health programs through school initiatives proudly launched its Early Intervention and Child Development Center at Sri Ramakrishna Central School. The Center was inaugurated by Padmashri Dr. R.V. Ramani, Founder & Managing Trustee, Sankara Eye Foundation in the presence of Shri. R. Sundar Joint Managing Trustee, SNR Sons Charitable Trust, Ms. Swathy Rohit, Chief Operating Officer, SNR Sons Charitable Trust and Mr. K. Gowthaman, President, Play School Owners Association, Coimbatore. This state-of-the-art facility is designed to support children with mild intellectual disabilities and learning difficulties, ensuring they receive the specialized care and attention they need for holistic development.

Sri Ramakrishna Hospital

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