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# **Paediatric Liver diseases**

The liver is one of the lar- cate liver disease and you damage gest organs in the body. It fills the upper right side of the abdomen inside the rib cage. The liver has many important functions, including filtering harmful substances from the blood so they can be passed from the body in stools and urine, making bile to help digest fats from food, storing glycogen (sugar), which the body uses for energy.

Baby Jaundice: Jaundice is very common in new born babies. Jaundice reaches its peak about at about 4 days of life and then gradually disappears in most babies by the time they are two weeks old. Jaundice does not necessarily mean your baby is ill. Jaundice is caused by your baby having an increased level of bilirubin in their blood. It is important that jaundice is monitored to ensure that the bilirubin level does not get too high.

Prolonged Jaundice: Sometimes jaundice continues after the baby is 14 days old in a full-term baby and 21 days in a premature baby. It is vital that the following is carried out:

Check the colour of your baby's stool: The stool of a breast fed baby should be green/daffodil yellow colour, the stool of a bottle fed baby should be green/English mustard colour. If your baby's stool is pale looks pale or chalky you must report this to your midwife or health visitor immediately. It could be a sign of liver disease.

baby's urine: The urine of a doctors treat it with the folnewly born baby should be colourless. If your baby's urine is yellow and/or the stool is pale, this can indi-

must report this to your mid- • Treating symptoms and wife, health visitor or doctor. Do not wait until after 14 days if you notice this before.

## What needs to be done?

It is important that your baby has a blood test called a split bilirubin blood test. This test measures the ratio of the conjugated and unconjugated bilirubin levels in your baby's blood. If the conjugated fraction is greater than 20% of the total bilirubin it indicates liver disease. Your baby should be referred to a specialist paediatric liver unit for further investigation.

# Cirrhosis in Children:

Cirrhosis in paediatric patients causes normal areas of the liver to be surrounded by scarred areas that do not function properly. Cirrhosis in children often stems from a wide variety of liver disorders, including Hepatitis B and Hepatitis C.

Cirrhosis Diagnosis

If your child's doctor suspects that your child has cirrhosis, he or she will perform tests to confirm or rule out the diagnosis. Tests may include:

Blood Tests - To assess how well the liver is working and determine a cause.

CT scan, Ultrasound, MRI Scan - To identify changes in the liver

Liver Biopsy – analyzing a sample of liver tissue removed via a thin needle inserted into the liver

# **Cirrhosis Treatment**

In general, cirrhosis can-Check the colour of your not be cured or reversed, lowing goals:

- · Controlling the cause of
- the liver damage Preventing additional

- complications
- Treating underlying medical conditions Your child's doctor may

prescribe drugs to treat the underlying cause of the liver disease. Other medications may be used to control symptoms or fight infections.

Some medications are prescribed to get rid of excess fluid in the body or reduce the risk of a blood vessel breaking. Others help your child's body cut down on its absorption of harmful waste products or toxins. If the complications of cirrhosis can no longer be controlled, or if the liver is in danger of no longer functioning, a liver transplant is often the best option. Many of the liver disorders that cause cirrhosis in children are not preventable, but there are precautions you can take. Make sure your child receives all

and hepatitis vaccines at the times your paediatrician recommends. If your child needs to take medications that may damage the liver, follow your doctor's recommendations

recommended immuniza-

tions including influenza

about blood tests. Balanced nutritional intake is important for people who already have cirrhosis of the liver can prevent or slow further liver damage by following their doctor's instructions regarding diet. Your child may need extra calories to grow properly and to maintain adequate overall strength.

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If the cirrhosis is more advanced and compromises the liver's ability to process protein properly, the doctor may recommend limiting protein. The doctor may also recommend limiting salt in your child's diet, because salt tends to make the body retain water.

They may also advise avoiding raw seafood. Make sure your child takes any vitamin supplements prescribed. Due to increased risk of infections, doctors recommend vaccines against flu, pneumonia, and hepatitis for people with cirrhosis.

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Hip pain Buttock pain Leg pain

Knee pain

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# FERTILITY OUTCOME IN ENDOMETRIOSIS



Endometriosis remains a top cause of infertility in females, both primary and secondary Infertile women are 6-8 times more likely to have endometriosis than fertile women. Chances of natural conception in women with different stages of the disease are: • Stage I and II - 30-35 % • Stage III - 10-15 % • Stage IV - less than 5%

### There are many ways in which endometriosis may interfere with fertility:

- 1. Mechanical interference, such as adhesions which can prevent egg pick up and transport Endometriomas- may prevent ovulation- make natural conception impossible.
- 2. Hostile and inflammatory intraperitoneal environment may impair ovulation, egg capture and quality, tubal function.
- 3. Cellular changes induced by the disease, increased oxidative stress levels which may lead to epigenetic changes.
- 4. Painful intercourse in cases of vaginal endometriosis- not a good environment for
- 5. Women with endometriosis may have coexisting endocrine and/or ovulatory disorders,
- including luteinized unruptured follicle syndrome, impaired folliculogenesis, luteal phase defect, and premature or multiple luteinizing hormone (LH) surge.
- 6. One of the concerns about treating patients with ovarian endometriomas is about the ovarian reserve - the number of healthy eggs you have - may be affected by excising the endometriomas.
- 7. Adenomyosis is another association seen in 20–30-% of patients with endometriosis. Adenomyosis reduces the chances of implantation and increases miscarriage rates.

# Treatment options available:

- 1. Stage 1 to 2 endometriosis: simple treatments like ovulation induction/IUI can be tried.
- 2. Stage 3 and 4 have very low success with IUI, IVF gives better success.
- 3. Surgical excision: only if the size of the ovarian endometriotic cyst is large and inaccessible
- 4. Specific IVF protocols are used while treating endometriosis/adenomyosis patients. Multidisciplinary and collaborative approaches to endometriosis-related infertility are the keys towards achieving the best outcomes.

A fertility team, which usually consists of reproductive endocrinologists, laparoscopic surgeons, embryologists, clinical nurses, counsellors, etc, form a vital part in achieving the best results. Despite the many options that exist, there is no simple answer or 'one size fits all approach and each case must be individualized.



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hronic kidney disease (CKD) in the 12th most common cause of death and 17th most common cause of disability. Prevalence of chronic kidney disease (CKD) in India is variable with regional differences at 4% to 17.2%. Incidence of CKD in India is estimated at 232/million population. True burden of CKD is not known in India because of lack of registry, lack of universal access to dialysis or transplantation and few dedicated centers of care. There are more than 130000 patients receiving dialysis in India of which hemodialysis constitute major form.

Patients with chronic kidney disease (CKD) who have progressed to end stage kidney disease (ESKD) require treatment in the form of dialysis or transplantation. Dialysis is of two types namely Hemodialysis and Peritoneal dialysis.

In hemodialysis patients' blood is dialyzed (filtered) of the waste products which are accumulated because of renal failure. For this procedure there is a need of access (arteriovenous fistula) from which blood is drawn, filtered and returned using needles and tubes. In the other type of dialysis called peritoneal dialysis the abdominal cavity is used for dialysis. In this type, a tube is placed into the abdominal cavity and it is left in that place permanently. In peritoneal dialysis fluid is filled into the abdomen via the tube which was placed in the abdomen and left for the amount of time prescribed by the Nephrologist. Filtering of waste products from the blood into this dialysis fluid takes place in the abdomen and after a said amount of time the fluid is drained outside. Peritoneal dialysis or hemodialysis both are only supportive management strategies for kidney failure. They are not curative as they do not replace all the functions of normal human kidney and the filtering capacity of these procedures of waste products is way below compared to normal kidney. Hemodialysis and peritoneal dialysis

carry their own set of issues and complications. Kidney transplantation is an alternative option to dialysis for ESKD patients. In Kidney transplantation a new kidney is placed in the body of the patient. As compared to dialysis in transplantation, it is a functioning kidney which is placed and it can do all the functions of normal kidney. transplantation is way forward in improving the condition of patients

### and make him dialysis free but transplantation also has its own set of complications.

Kidney transplantation is of two types based on the types of donor. They are Deceased donor transplantation and Live donor transplantation.

First is from the deceased donor in which kidneys are taken from brain dead patients after appropriate sions and tests. donor transplantation patient needs to register themselves in their respective state organ registry. After a waiting period, once the patient turn comes, he will be transplanted. The other form of transplantation is the live donor transplantation in which a healthy person donates a single kidney of his / her to a patient of end stage kidney failure. Live kidney donation is the major type in India and gradually cadaveric donation is picking up especially in Telangana because of robust Jeevandan program. Hospital data has shown that 61% of stage 5 CKD patients were not offered any form of dialysis therapy or transplantation. Remaining 32% were on dialysis, 5% on peritoneal dialysis and only 2% received a kidney transplantation.

# Disadvantages

- · Before a fistula is made, patient may require a dialysis catheter placed in the neck which can lead to
- muscle cramps and lood loss etc.
- Low blood pressure can lead to abdominal cramps, nausea, vomiting, light headedness.
- Loss of work hours for dialysis treatment and travel to dialysis
- · Done at home with no need to travel to dialysis center
- proper precautions

# Disadvantages

- in proper way
- · Risk of infection of abdominal
- · Increased risk of hernia (weakening

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- Common complications related to dialysis procedure include low blood pressure (hypotension), high blood pressure (hypertension, low blood sugars (hypoglycemia),
- Peritoneal Dialysis:

# Advantages

- Can be done at work place with
- Need to learn to use the equipment
- cavity (peritonitis)

- · Risk of rejection of transplanted organ and loss of its function
- Risk of infections and rarely cancers because of immunosuppressive medications

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If we compare the quality of life on dialysis and after transplantation studies have clearly shown better quality of life with transplant unequivocally.

"Quantity of life (survival) is also better in patients of kidney transplantation compared to remaining on dialysis."

are transplanted, they survive better than those remaining on dialysis. The difference is much bigger in younger patients in terms of survival.

Even if we take elderly people who

If we compare the risk of death between staying on dialysis and after kidney transplantation it is greater in transplant recipients for around first 3 months. After three months the risk of death reverses and equals the hemodialysis by 6-9 months and there



**Even though living kidney donation** multitude health problems in him later is best in terms of outcome the deceased donor transplantation is many times better than remaining on dialysis. Early transplantation once the patient

lands in ESKD is helpful to patients rather than remaining on dialysis for some time and then undergoing transplantation. The duration of survival after transplantation is greater in patients who undergo transplantation early compared to patients who remain on longer periods of dialysis before transplant.

Not all patients with end stage kidney disease can be transplanted because of various contraindications already mentioned. In older age and severe vascular or heart disease patients it is better to treat with dialysis rather than undergoing kidney transplantation

in life. People with hypertension which is adequately controlled on single medication with no other damages in the body due to hypertension can donate. Donors kidneys are evaluated by urine examination, blood tests, imaging studies (ultrasound and computed tomography) and nuclear studies to assess their structure and function. If there is any abnormality in the tests done for kidney function which is reconfirmed that donor is denied from donation. Donor needs to understand the procedure of donation including the hospital stay, associated complications of surgery and possible long-term risks associated with donation. There should not be any monetary reason for donation and that needs to be ruled out before proceeding with surgery.

Absolute contraindications

The following are the absolute

contraindications for kidney donation

· Hypertension on multiple

· Obesity with body mass index of

greater than 35 with other

Pre existing renal and familial

Current cancer or treated cancer

with significant risk of recurrence

· Infections like Hepatitis B, HIV,

risk factors for heart disease

ssociated health issues

· Recurrent kidney stone disease

medications with additional strong

for Kidney donation

· Age less than 18 years

Diabetes mellitus

kidney disease

· Heart disease

Hepatitis Cetc.

· Active substance abuse



If we take the risk of end stage kidney disease (ESKD) after kidney donations, studies done in western countries have shown that life time risk of ESKD in persons who have donated is 0.9%. If we consider the risk in healthy people who do not donate it is 0.14%. So, there is an increase in risk of kidney disease over life time in donors but the absolute increase is itself very

Women who donated have higher risk of gestational hypertension or pre-eclampsiabut there is no difference in outcomes such as requirement of cesarean section, post-delivery bleeding, preterm birth (delivery at <37 weeks of pregnancy) or low birth weight babies (<2500gm).

· Donors who are obese have 2-fold higher risk of end stage kidney disease (ESKD) over 20 years after donation There may be increased risk of

hypertension and gout in donors.



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# KIDNEY TRANSPLANTATION Best treatment for kidney failure patients

### Advantages of Living donor transplantation compared to deceased donor transplantation

- Less waiting time for transplantation
- · Longer survival of transplanted organ as they are taken from health donors who are screened and
- scheduled surgery · Better patient survival
- · Less immunosuppressive drugs requirement Less morbidity

Absolute contraindications for

- Kidney transplantation · Health condition that could lead to
- death early (<1-2 years) · Recently treated or active cancer
- · Poorly controlled mental illness
- · Current drug or alcohol dependence · Severe dementia · Poor compliance to treatments or

# Kidney transplantation vs Dialysis: what to choose?

Dialysis (Hemodialysis or peritoneal dialysis) as an option compared to kidney transplantation fares less overall

## Hemodialysis: Advantages

medications

Requires few hours of treatment that too not everyday





of the abdominal muscles) from the pressure of the fluid inside the abdominal cavity

· Other complications include weight gain, abnormal lipid levels and high blood sugars etc.

### Kidney transplant Advantages

# · Better control of symptoms of kidney failure with almost full

- No need of regular dialysis · Better quality of life · Long life expectancy compared to
- Disadvantages

dialysis

- · Involves a major surgical procedure with risks during and after surgery like bleeding, infection and very rarely death
- · Required to take medications life long without fail

# DISEASED



after it favors transplant with transplant patients having greater survival compared to patients remaining on hemodialysis over long

### Because of the above-mentioned reasons, a patient with ESKD should be first assessed for their suitability for transplantation.

The best first option is a pre-emptive transplant from a living related donor or living donor. Pre-emptive means undergoing transplant even before starting dialysis. The advantage of this is that there will be no need for initial period of dialysis and the creation of vascular access (arteriovenous fistula). It also enables proper planning of transplantation with a proper matched individual which will have impact on long term survival of transplanted organ and patient. If there is no living donor available for transplantation then next best option is deceased donor transplantation.

contraindications for transplantation dialysis is the only option to pursue. Live Kidney donation

because of the risks associated with

transplantation. In the patients with

# There are set guidelines and rules in

India with regards to transplantation of organs called transplantation of human organs and tissue rules, 2014 from government of India. Live donation can be from near relative or other than near relative. Near relatives include spouse, son, daughter, father, mother, brother, sister, grandfather, grandmother, grandson and granddaughter. Any other person falls into the other than near relative category. Donation from near relative cases is authorized by the hospital conducting the transplant. Donation from other than near relative requires a scrutiny to prevent selling of organs and is permitted only by authorization committee of district/state after proper scrutiny.

Once the donor is selected whether from near relatives or an authorized donor (who is not a near relative), they are evaluated for fitness to donate Donors undergo a range of blood and imaging tests to assess the fitness for donation. Evaluation includes ruling out the presence of diseases in the donor which may later affect the kidney later in the life like diabetes, hypertension, kidney diseases, infections etc. If donor is young and diabetic, he is not deemed to be fit for donation as he has long life expectancy and diabetes can lead to

# Risks of Living kidney donation: what do we need to know?

- · Risks related to surgery (removal of single kidney) like blood loss, infections (urinary infections, lung infections, wound infection at surgical site), wound complications like hernia and rarely death
- There is no difference in the longterm risk of death when we compare donors who have donated their kidney and non-donors

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