What is the liver?

A normal healthy liver is reddish brown in colour. It lies on the right side of the abdomen beneath the lungs. It is divided into two major lobes called the left and right lobes. The lobes are different sizes and shapes. These lobes are separated by a band of tissue called the falciform ligament.
What does the liver do?

It’s complicated! Some people think of the liver as the body’s power station which supplies energy to the body. It is also a chemical plant and a waste station. All the functions of the liver involve processing blood in some way. The liver breaks down the nutrients and chemicals in your blood. It stores the nutrients your body needs until your body needs them. When your body needs them it releases them into the blood again. The liver produces substances such as proteins needed to keep your body functioning. It also helps get rid of the waste products the body no longer needs.

There are over 500 vital functions that your liver does every day. These functions come under 3 headings:

1. **Production and transportation of bile.**
   Bile is a yellow/green thick liquid made by your liver. Bile is carried to the bowel via the biliary system. One role of bile is to remove waste products such as bilirubin and toxic chemicals from the liver. Bile also contains useful things which help us absorb fat from our food. Some bile substances (e.g. bile salts) are returned from the bowel back to the liver for re-use. So even though bile is partly made up of waste products, it certainly doesn’t go to waste!

2. **Storage and conversion of nutrients for energy.**
   Fats, carbohydrates and proteins are all ways we can store energy. The liver stores them and changes them into energy as we need them.

3. **Production of proteins.**
   Proteins are very important for normal body function. Proteins are used for fluid balance, blood clotting & immunity. They are also used for balancing hormones and for growth and nutrition.

The liver has a remarkable ability to renew itself.
What is portal hypertension?

This is an important complication of liver disease which is closely monitored pre-transplant. Portal hypertension is a common problem in children with liver disease. As the liver becomes increasingly scarred, it gets stiffer and it is difficult for blood to flow through it. Blood has to make its way back to the heart through other ways. Increasing amounts of blood have to flow through veins that are not designed to cope with high volumes.

What normally happens?

The heart pumps blood to the intestine where the nutrients from food are picked up. The energy laden blood is then pumped to the liver for processing. The blood then drains back to the heart.

What is portal hypertension?

- Portal = the blood vessels that drain the stomach and intestine
- Hyper = high
- Tension = pressure

Portal hypertension is high blood pressure in the blood vessels draining the stomach and intestine.

Portal hypertension is caused by something stopping the normal flow of blood through the portal vessels. This can happen before, within or after the liver.

Why does my child have portal hypertension?

My child has a normal liver why does he/she have portal hypertension?

Blockages before the liver are called pre hepatic. They are the most common cause in children who do not have liver disease. The blockage can be caused by narrowing of a blood vessel called a portal vessel or by a blood clot blocking the flow. It can also be caused by blood vessels not being formed properly. It is this increase in blood flow through the alternative routes that is called portal hypertension. Portal hypertension is managed in the same way regardless of what is causing it.

Why does my child with liver disease have portal hypertension?

In liver disease the scarring in the liver makes it hard for blood to pass through it. The blood flows back to the heart via different paths. It is this increase in blood flow through the alternative routes that is called portal hypertension. Portal hypertension is managed in the same way regardless of what is causing it.
What problems does portal hypertension cause?

- Varices - varicose veins in the oesophagus (tube between the mouth and stomach) & intestine
- Increased spleen size and function
- Ascites - a collection of fluid in the free space in the abdomen. It is caused by swelling of the blood vessels in the intestine and fluid leaking out from the blood stream.

How will my doctor know if my child has portal hypertension?

If the spleen is larger than normal, this suggests portal hypertension. The spleen size can be measured by the doctor examining your child’s abdomen or by ultrasound scan.

Blood tests: A low platelet count or white cell count may suggest portal hypertension.
What are varices?

Varices are “varicose veins” of the oesophagus and intestine. When blood finds it difficult to go through the liver, it makes it’s way back to the heart through alternative blood vessels. These blood vessels are not always able to cope with the increase in blood flow. The higher flow increases the pressure in the veins. This makes the veins swell and some of the veins have thin walls. This leads to a risk of the vein bursting which may cause serious, or life-threatening bleeding.

Where can bleeding occur?

The common places that bleeding can occur from are the oesophagus, stomach, and from the rectum (bottom).

How will I know if my child has a bleed?

- Vomiting blood
  Blood in the vomit may be fresh which is bright red. It can be old blood which may be in clots, look black, or like coffee ground.

- Passing blood in their stool
  Blood in the stool is usually old and causes the stool to be black, tarry and strong smelling. However, bright red fresh blood may also be passed.

Note: this is different from the common symptom of flecks of bright red blood around stools that might occur from constipation.

- Your child becoming very pale or fainting

What do I do if my child bleeds?

CALL 111 AND ASK FOR AN AMBULANCE

- Tell the Ambulance Operator what has happened and that your child has portal hypertension and liver disease.
- Take anything that might help confirm that there was bleeding (bowl, blood stained clothing etc)

At the hospital your child will be assessed and stabilised by the emergency team. There are a number of things which may need to happen when a bleed has occurred these include:

- Having an IV put in
- Starting special medicines via the vein
- Looking at the intestine with a camera, which is called an endoscopy

What can be done to prevent bleeding?

If your child has signs of portal hypertension an endoscopy may be performed.

Your child may also be put on medicine to make it less likely that they will bleed.

If you are outside Auckland, your child will be stabilised in the local hospital and transferred to Starship as soon as possible.
What is an endoscopy?

The Dr will be using a special camera to see the walls of the Oesophagus. The walls of your child’s stomach and the first part of the bowel are also important and will be checked carefully. Your child will be asleep while this is happening.

The doctor will be looking for blood vessels which are swollen and sticking out from the normal walls. These can sometimes look like bunches of grapes or varicose veins. Sometimes the walls of the stomach might be red and broken like a new graze. Both these things tell us there is higher blood flow in vessels than normal. The Dr will also be looking for signs of recent bleeding. This might be scabs from sites which have healed or old blood in the stomach.

What happens if there are Varices?

This depends on where they are, how big they are and if your child has a history of bleeding.

There are several things the doctor can do to block off the varice which might cause a problem with bleeding.

Bandining is used the most often. This involves placing a rubber band around the vessel to block it off.

If your child is too small to get the equipment down to band then a medication will be injected which causes a scar in the varix. An injection of glue can also be used in some cases. Your doctor will tell you what the best option is for your child.

What happens if the Varices come back?

If the cause of the portal hypertension has not been fixed then varices do come back. Until the problem causing the hypertension is fixed the Dr will book repeat endoscopy regularly to keep watching for new varices and treat them as needed. This may be every few weeks or few months depending on your child. This helps reduce the chances of bleeding but does not stop it completely.

If you are outside Auckland, your child will be stabilised in the local hospital and transferred to Starship as soon as possible.
What is Ascites?

Ascites is extra fluid inside the abdomen, but outside of the intestine. Ascites develops when the liver is unable to make enough protein.

How do I know if my child has ascites?

If your child’s abdomen is getting larger, then they may have ascites. This is more likely if it happens quickly over a few days or couple of weeks.

Your doctor may be able to tell by feeling your child’s abdomen or by an ultrasound scan.

How is ascites treated?

Ascites is usually treated with special medicines to help with extra fluid. As liver disease gets worse a combination of medicines and Albumin infusions might be needed.

Can ascites get infected?

Yes - the fluid can get infected, this infection is called peritonitis. If your child has an episode of peritonitis while waiting for transplant they will be suspended from the list until the infection has been fully treated.

How will I know if my child has peritonitis?

If your child has the following symptoms please bring him her to the hospital immediately.

The usual signs of peritonitis are:

- A rapid increase in the size of their abdomen.
- Pain in the abdomen.
- Fever.
- Diarrhoea or vomiting.

Peritonitis is treated with IV antibiotics.

Key points to remember

Portal hypertension is an important problem and bleeds can be life threatening. Things to look out for are:

- Bleeding - If your child ever vomits fresh or old blood, or passes stool which contains fresh or old they are likely to be bleeding. Your child needs to go straight to hospital - if in doubt call an ambulance.
- Remember fresh blood is red and old blood can be coffee or black coloured.
- Abdomen getting larger.

If your child’s abdomen becomes suddenly larger then they need to be seen by a doctor that day. This is especially important if they are also looking unwell.
## Important telephone numbers

<table>
<thead>
<tr>
<th>Service</th>
<th>Contact Information</th>
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</thead>
<tbody>
<tr>
<td>Starship (Auckland City Hospital)</td>
<td>09 307 4949</td>
</tr>
<tr>
<td>Liver Nurse Specialist</td>
<td>Cate Fraser-Irwin &amp; Karyn Sanson</td>
</tr>
<tr>
<td></td>
<td>Oncall phone - held by the nurse responsible for outpatients 021837870</td>
</tr>
<tr>
<td>Gastroenterology Consultants</td>
<td>Care of Dani (Team Support) Extn 22292</td>
</tr>
<tr>
<td></td>
<td>Out of business hours please contact On-call Consultant via Auckland City Hospital switchboard 09 307 4949</td>
</tr>
<tr>
<td>Team Support</td>
<td>09 3074949 Extn 22292</td>
</tr>
<tr>
<td>Clinic bookings</td>
<td>Deborah Robb 09 307 4949 Extn 22490</td>
</tr>
<tr>
<td>Ward 26B Medical Specialties</td>
<td>09 307 4949 Extn 25760</td>
</tr>
</tbody>
</table>

### How do I contact the Clinical Nurse Specialist?

The Clinical Nurse Specialist team for Liver & Gastroenterology are often your first point of contact when you have questions about your child relating to their illness.

Our role is national so regardless of where you are living within New Zealand we may need to provide support or guidance from time to time. We work closely with the paediatric services (nursing and medical) in your home town. Most of our families have children with ongoing medical needs due to poor or deteriorating liver disease. Some have also had a liver transplant.

#### When contacting the team we do ask that you consider if your question is urgent or non-urgent.

Because your calls are important to us, we have launched an on-call phone so that the CNS team can be contacted quickly as needed.

**If Urgent contact:**
- CNS team during business hours 8- 4.30pm weekdays
- By Phone: 021 837 870
- Please **DO NOT TEXT MESSAGE** us as we have no way of knowing you have tried to contact us if the message is delayed or not delivered.

- After hours and weekends: take you child to your GP clinic or nearest emergency department for assessment
- If outside Auckland please first contact your local team for advice.

**If Non urgent:**
- We can be emailed: StarshipLiverCNS@adhb.govt.nz
- This email will be received by each CNS and replied to as appropriate.
- If you require new PRESCRIPTIONS please contact your GP or our team support:
- For questions relating to CLINIC VISITS please contact our clinic scheduler