A Booklet on
Cerebral
Ventricular Shunts

BC CHILDREN’S HOSPITAL
An agency of the Provincial Health Services Authority

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Introduction

This booklet addresses some of the general concerns of families whose children need shunts. Your child’s particular problems cannot be discussed in a booklet. No booklet can replace face-to-face discussion. Use what you learn here to help you discuss your child’s condition with greater understanding. Please use the back of this booklet for notes.

What is a shunt?

A shunt is a thin, flexible tube with a valve. It is positioned in the brain to drain excess fluids out of the head. The fluid is “shunted” elsewhere in the body where it is absorbed into the blood stream.

Why is a shunt necessary?

A shunt is necessary when too much cerebrospinal fluid (CSF) collects in the brain.

What is a CSF – cerebrospinal fluid?

Cerebrospinal fluid is a watery substance containing proteins, salts and sugars. It acts as a sort of water cushion to protect the brain from injuries. Also, it carries food to the brain and waste from the brain. Cerebrospinal fluid contributes to the right chemical environment for brain activity.

How is CSF formed?

CSF is formed within the brain itself. Most people imagine the brain as solid tissue. In fact, it contains 4 “cave-like” spaces called ventricles and space around it called subarachnoid space.

CSF is formed constantly in the lateral ventricles (one on the left and on the right). It is formed from fluid filtering out of the blood. The brain and spinal cord are bathed in 1/2 to 2/3 cup of this fluid all the time. But, about 4 times this amount is produced each day. The extra is absorbed back into the blood. Therefore, any blockage quickly results in a build up of fluid which overfills the spaces within and around the brain.
How does CSF circulate?

CSF passes from the right and left lateral ventricles into the third ventricle (below them) through a narrow connecting channel called a **Foramen of Monro**.

An even narrower channel called the **Aqueduct of Sylvius** carries the fluid from the 3rd ventricle into the 4th ventricle (on the floor of the brain). From there it flows, through 3 small openings, around the spinal cord. CSF also passes over the surface of the brain in the subarachnoid space before being absorbed by the blood through special tissues connected to a large vein on the surface of the brain.

![Diagram of the brain showing the flow of CSF](image)

What causes the abnormal build up of CSF?

The build up is usually caused by a blockage in the system through which the fluid circulates. If the fluid cannot circulate properly it collects in the spaces in the brain and more and more pressure builds within the brain and skull (intracranial pressure). The build up of large quantities of fluid leads to the condition called hydrocephalus (hydro – water; cephalus – head). This condition must be treated quickly to avoid serious problems.

What causes the blockage?

A blockage can result from a number of different causes. The most common kind of blockage is a narrowing (**stenosis**) of closing (**occlusion**) in one of the channels which link the ventricles. This may be an inherited or unexplained malformation or it may occur because an infection or bleed has scarred tissues in the brain. An abnormal growth may also block the flow.
The blockage is sometimes in the subarachnoid space rather than the ventricles. Such a blockage prevents the absorption of CSF into the bloodstream. This type of blockage is usually the result of a bleed, meningitis, a tumour or cyst.

The shunt provides a way for the cerebrospinal fluid (CSF) to bypass the blockage. If fluid builds up pressure, the valve of the shunt opens. The valve shuts when the pressure goes down to normal levels.

**Why does the doctor suspect that my child has a blockage?**

The first sign of hydrocephalus may be rapid increase in head size. An ultrasound scan shows this before birth. The other signs of pressure building within the skull (increased intracranial pressure) vary depending on age.

**Newborn – 2 year olds:** Bulging, tense fontanelle (soft spot), eyes which seem to look down and move at random, unusual irritability, vomiting, seizures and high pitched crying are signs of increased intracranial pressure.

**Older Children:** Nausea, vomiting, morning headaches, bulging downcast (sunsetting) eyes, tiredness and personality changes may suggest a problem.

**How does the doctor know that my child has a blockage?**

The doctor needs a picture of the ventricles. The picture may also show the position of a blockage. There are different ways to obtain this picture.

**CT Scan**

The CT Scan is the one most commonly used. This is a computerized X-ray examination. The pictures shown here were taken in this way.

A CT Scan is painless but the child must remain perfectly still. Some young children can’t be still and need to be sedated.

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**CT Scan showing hydrocephalus**

**CT Scan after shunting**
Ultrasound

In very young children whose fontanelle has not yet closed, an ultrasound is usually preferred. Ultrasound uses sound waves to create an image of the brain tissue. It is a safe, painless test. The sound waves cannot pass through bone, therefore, the test can only be used while the fontanelle is open. The picture below was taken with an ultrasound.

Ask for a more detailed explanation if your child is having this test.

Ultrasound showing hydrocephalus

MRI

A magnetic resonance image (MRI) is created by a magnet along with radio waves. An MRI picture can show many different views of the brain as well as the flow of the CSF. With these pictures may be possible to find the blockage.

Does hydrocephalus affect a child’s intelligence or other abilities?

Hydrocephalus itself does not damage the brain if the pressure is relieved quickly. Sometimes the same problem that has caused the hydrocephalus has damaged the brain. Your doctor will discuss your child’s particular condition with you.
Is there any alternative to a shunt?

For most children with hydrocephalus, a shunt is the best and only treatment. There are a few situations in which there are other options:

- When a tumour is causing the blockage it is sometimes possible to remove the tumour so that CSF again flows normally.
- When the blockage is in the aqueduct of sylvius, it may be possible to make a small opening between the 3rd ventricle and the subarachnoid space.

The surgeon makes a small hole in the skull beside the fontanelle. He gently pushes a special tube-like instrument called an endoscope into the 3rd ventricle. The tube has a light and a separate channel through which liquid or air can flow. Once the tube is in position, a small hole is “blown” into the base of the 3rd ventricle.

This provides an outlet for the CSF from the ventricles.

- When the blockage is from a bleed in the brain, a temporary shunt may be put in. This is called a ventricular reservoir. It is used mostly with infants. Because the skull is still soft, it is fairly easy to push a small tube through it and into a ventricle. The tube is attached to a container (about the size of a dollar). The container is covered by a flap of skin. The CSF can overflow into this container and be pulled out with a syringe. This drains off CSF without touching the brain until the circulation returns to normal.

What does a shunt look like?

There are several different types of shunts.

Most have 3 parts:

1. a small piece of tubing (ventricular catheter)
2. attached to a valve inside a reservoir (dome/bubble). The valve connects on the other end to
3. a long length of narrow tubing (peritoneal catheter).
The catheter is positioned in one of the lateral ventricles and the valve sits on the skull. The tubing runs under the skin down the side of the head and neck along the chest or back and ends in the abdominal cavity. The CSF which is shunted into the abdominal cavity is absorbed back into the bloodstream.

Sometimes the surgeon places the end of the tubing into one of the large veins at the base of the neck. These veins empty directly into the heart.

**How is the shunt put in place?**

The shunt is “implanted” in the operating room while your child is under a general anesthetic.

1. Your child is prepared for this surgery as for any other.

2. You wash the hair and scalp with a special disinfectant soap. The surgeon may want a patch of hair shaved. If so, this is done in the operating room. If you or your child prefer all the hair shaved, tell the nurse.

3. The surgeon makes a small cut on the scalp. A tiny hole is created in the bone through which the catheter is guided into a lateral ventricle.

4. The valve is connected to the catheter and the tubing.

5. Another small cut is made at the point where the tubing with end – usually the abdominal cavity.

6. Using a little metal rod, the surgeon prepares a tunnel just under the skin between the cut on the scalp and the one on the abdomen. Sometimes a few more cuts are needed as points between. The long length of tubing is then threaded through the tunnel and attached at one end to the valve and the other is set into the abdominal cavity. The tubing is long enough to allow the child to grow to full height without another surgery.

**Putting a shunt in place:**

Implanting usually takes about 2 hours. The child stays another 2-3 hours in the Recovery Room before returning to the unit.

**How long will my child be in hospital?**

Most children are well enough to go home a day or two after the surgery. A few may need up to 10 days in hospital. The stitches are normally removed on about the 7th day. This can be done by your family doctor.

**After the shunt is in, will my child’s head return to a normal size?**

If the shunt works well, the head may get smaller or stay the same size while the rest of the body grows. Unless the head was very large before shunting, it will come to match the body size.

Adolescents with hydrocephalus, who were treated when they were very young, have normal size heads.
How long must the shunt remain in my child’s head?

The short answer is “for life”. Shunts are not removed unless they cause problems. A few children become “shunt independent”. They seem to develop alternative paths along which the CSF can flow. It is hard to know to which children this happens or how it happens. It is best to assume that your child will always need a shunt.

Will the surgery damage brain cells and affect intelligence?
The catheter goes through the brain in a way that does not affect your child’s natural abilities.

LIVING WITH A SHUNT:

Will my child need special care after the surgery?

Some children recover quickly – others take longer. It depends on your child’s age, general health and whether there are setbacks.

For the first week or two make sure your child gets enough rest. But, if your child feels well and wants to get going, allow it. You know your child best and will be able to judge when he needs to slow down. Your child can shampoo when stitches are out.

Arrange to see your family doctor one week after discharge from the hospital.

The neurosurgeon will want to see your child in 2 months. Sometimes a CT or ultrasound scan is done at this follow up visit.

*If your child seems ill or shows any unusual or irritable behaviour, call your family doctor immediately. Complications with the shunt can be serious. They need urgent attention.*

Will living with the shunt be difficult for my child?

The shunt will not break easily. It can stand up to the bumps and jumps of an active child who climbs, rolls and stands on her head. The skin covering the shunt will keep the dirt and germs out once the scar is healed. In other words, your child is able to live a normal life with a shunt. The only exception may be rough sports like rugby or ice hockey. Speak to the doctor if this is an issue for your child.

All children need to play and experience the world as fully as possible. Don’t overprotect your child or regard him as sick. Set standards and expectations as you would for any other child.
PROBLEMS WITH SHUNTS

1. Malfunction

Shunts are mechanical objects. They can malfunction.

**Shunt blocks:**

It may become plugged with body cells or tissue growing around the open ends of the tubing.

**Shunt only works on and off or works, but not well enough:**

This type of malfunction can go on for a long time before it is recognized. The symptoms may be different from what one expects with malfunction. The child may seem clumsy, trip, complain or not seeing well, wet himself, do poorly at school or have a change of personality.

**Tubing breaks or disconnects:**

This happens when scar tissue grows around a section of the tube. The scar holds that piece of tube while the tubing above and below moves. The movement wears the tubing at this point and it eventually breaks. The loose piece of tubing usually moves into the abdominal cavity. It seldom causes a problem but the spinal fluid may leak into the tissues causing swelling.

*If you notice a soft swelling anywhere along the shunt line below the scalp scar, you should have this checked by your doctor.*

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**SOME SIGNS OF A SHUNT MALFUNCTION**

<table>
<thead>
<tr>
<th>Infants</th>
<th>Toddlers</th>
<th>School Children</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bulging fontanelle</td>
<td>Headaches</td>
<td>Headaches</td>
</tr>
<tr>
<td>Vomiting</td>
<td>Vomiting</td>
<td>Vomiting</td>
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<tr>
<td>Poor Appetite</td>
<td>Poor Appetite</td>
<td>Poor Appetite</td>
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<tr>
<td>Sleepiness</td>
<td>Lethargy</td>
<td>Lethargy</td>
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<tr>
<td>Irritability</td>
<td>Irritability</td>
<td>Irritability</td>
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<tr>
<td>Seizures</td>
<td>Seizures</td>
<td>Seizures</td>
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<tr>
<td>Swollen/red shunt tract</td>
<td>Swollen/red shunt tract</td>
<td>Swollen/red shunt tract</td>
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<tr>
<td>Sunsetting eyes</td>
<td></td>
<td>Drop in school performance</td>
</tr>
</tbody>
</table>
How is the malfunctioning shunt recognized?

You and your child (as he gets older) should know the usual signs of shunt complications. See a doctor at once if you suspect a problem.

A child with a shunt malfunction may have all or only one or two of the signs listed in the table. Some children may have clear symptoms and others mild changes. It may be hard to know if the shunt is malfunctioning, or if the child is teething or coming down with a cold. Your doctor knows it’s hard for a parent to decide if this is serious. So, when in doubt, call your doctor for advice. Your sense that all is not well is important information for the doctor.

If the shunt malfunctions once, it is likely that you will recognize the signs if it happens again. The signs seem to be the same each time.

Your doctor will need to check the shunt.

- A CT Scan, MRI or ultrasound can be used to check the size of the ventricles. If the shunt is malfunctioning, they will be larger than in the pictures taken while the shunt was working.
- A series of ordinary x-rays (shunt series) will show a break or disconnection in the tubing.
- In a few situations the ventricles do not enlarge yet the child has signs of shunt malfunction. The doctor may ask for a test called a shunt scan. The test shows whether the CSF is flowing freely through the entire shunt. A tiny amount of radioactive substance (radioisotope tracer) is injected into the reservoir of the shunt. A special camera then records the flow through the tubing. The pictures below show the images created by this test.

The test is done in the Department of Nuclear Medicine. It takes 45-60 minutes. To prevent infection, a patch of hair about the size of a nickel is shaved and the area thoroughly cleaned. The radiologist and technician wear masks.

The child will feel the needle prick briefly and may “feel funny” because of the change of pressure when the substance is injected. You may stay with your child, after the injection, if you wish.
Treatment of malfunction:

The treatment depends on the degree of malfunction and its effect on your child. If the shunt malfunctions only now and then, and the child is well, you and the doctor may decide to watch more closely but not treat. If, however, the tests show that the ventricles are much enlarged, and your child is not well, treatment is urgent. The shunt is removed while your child is under an anesthetic. It is repaired or replaced.

This is called a shunt revision. It is similar to the procedure for placing the shunt.

Shunt revisions have a risk that is not there when the shunt is first put in. Pulling out the shunt can cause a bleed in the brain which damages some brain tissue. Your physician will discuss this concern with you.

2. Shunt Infection

Tubes put into the body can become infected. Germs may breed in or on the tubing and cause infection. The signs of an infected shunt are like those of a shunt malfunction. With an infection, your child may have a fever. Sometimes the fever is the only symptom.

*If your child has an unexplained fever which lasts longer than 24 hours, see your doctor.*

Treatment of infection:

Infection is a serious complication. It can happen at any time. There is a particular risk after the surgery, before healing is complete. Infection can occur in the shunt itself or in the wounds. In a few cases, infection in the shunt can be treated by running antibiotics through the shunt and into the blood. Usually, the infected shunt must be taken out. The child is then given antibiotics. Once the infection is cleared, a shunt is put back in. If the intracranial pressure builds too much without a shunt, an external drain or temporary shunt is put in.

**Note:** Germs can enter the body easily during dental work. Tell your doctor if your child needs work on the teeth or gums. The doctor may suggest an antibiotic to guard against the increased risk of infection.

3. Subdural hematoma (*bleeding on the surface of the brain*)

When a shunt is working well it may drain the CSF quickly. The ventricles shrink and the brain grows smaller. The delicate veins which run from the surface of the brain to the skull can become stretched and break. Small amounts of blood spill into the subarachnoid space. This type of bleed is less likely to cause damage than a bleed inside the brain. However, it must be treated or pooling blood creates another kind of pressure on the brain.

The signs will be the same as for a shunt malfunction but the cause is pressure from the bleed not CSF build up.

**Treatment:**

A small burr (hole) is made in the skull. A tube is placed to drain the blood and allow space for ventricles to fill. It may be necessary to change the valve on the shunt.
4. **Slit ventricle syndrome**

This problem may happen to the older child who has had a shunt for some time. The shunt pulls so much CSF out of the ventricles that the ventricle shrinks (like a balloon with too much air removed) and falls in over the end of the catheter blocking it. When the CSF over-fills the ventricle, it opens again. The problem seems to come and go. The symptoms are usually headaches and vomiting.

**Treatment:**

This condition is treated in one of two ways:

- The shunt valve is replaced with one which allows less CSF through. This requires another surgery.
- A small hole (2” in diameter) is made in the skull above and behind an ear. It is covered with a skin flap. The purpose is to allow space for the brain to swell or shrink a little without damage. It allows for a shunt which is not working perfectly. In time, new bone forms and the hole fills in.

5. **Inguinal hernias**

Newborns, especially premature boys, may develop a hernia as a result of a shunt. This may be because the CSF which empties into the abdominal cavity puts pressure on the muscle wall.

**Treatment:**

Hernias usually need a surgical repair.

**How common are complications?**

Some children have a shunt inserted as a newborn and are well from then on. In their early teens, they may need a longer shunt.

Other children have a malfunction or infection once or twice but, for the most part do really well. Unfortunately, there are few children who have problem after problem and require many revisions. Why this is so, we cannot say. It is impossible to predict what is ahead; it’s best to be positive.

Teach those people caring for your child such as the school nurse, teachers and babysitters about your child’s shunt. Ask them to tell you at once if they notice signs of malfunction.
What follow up care is required for the child with a shunt?

The neurosurgeon will schedule visits, usually four a year, until two years of age, and twice a year after that. It is important to keep these appointments. During these visits, your child’s head is measured, growth and development checked and other examinations may be done.

As well, the doctor may ask for x-rays to check on the amount of tubing left for growth and a CT scan to measure the size of the ventricles.

When and what should I tell my child about this disorder?

Children who have hydrocephalus requiring shunting have a life-long condition. They need to understand the condition and the reasons for the shunt just as they need to understand other parts of their body and its care.

Tell your child about it, a little at a time, as situations arise. For instance, after a shampoo, you may say, “Can you feel this little bump? It’s called a shunt. It has a special job to do to keep you well”.

You can add more and more information each time. For example, “There is watery stuff inside our heads which helps our brains to work. When there is too much of this water, there is not enough space for our brains. So we all have a little drain inside our heads through which the extra water can run out. Your little drain is blocked and that’s why the doctor has put this special part inside your head to drain the extra watery stuff from your head.”

These are only ideas. Follow your own feeling about what is right for your child. Of course, you should answer all your child’s questions in terms she can understand. But don’t wait for questions. Make times to talk with your child in an open, honest and matter-of-fact way. This shows that you are accepting of the condition. When you talk comfortably about the shunt, your child will be more willing to bring his concerns to you. Remember that children may need to have the same explanation repeated many times and again as a teenager.

If you have concerns which are not covered here, or would like to discuss any of these issues in more detail, speak with a health care professional caring for your child. At BC Children’s Hospital, we believe that parents are partners and should be as informed as possible about their child’s disorder.

The Family Resource Library has more information about Hydrocephalus and the tests described here. There is also advice on helping your child manage tests and other procedures.
**Glossary:**

<table>
<thead>
<tr>
<th>Term</th>
<th>Description</th>
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<tbody>
<tr>
<td><strong>Acquired hydrocephalus</strong></td>
<td>Hydrocephalus caused by something which happens to the brain after birth.</td>
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<tr>
<td><strong>Aqueductal stenosis</strong></td>
<td>A blockage in the channel that connects the third and fourth ventricles in the brain.</td>
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<tr>
<td><strong>Angiogram</strong></td>
<td>A diagnostic test that provides an x-ray image of the blood vessels.</td>
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<tr>
<td><strong>Catheter</strong></td>
<td>Flexible, hollow tubing.</td>
</tr>
<tr>
<td><strong>Choroid Plexus</strong></td>
<td>Tufts of tissue found in the ventricles which produce the cerebral spinal fluid.</td>
</tr>
<tr>
<td><strong>Communicating hydrocephalus</strong></td>
<td>Hydrocephalus caused by a blockage in the subarachnoid space rather than in the ventricles.</td>
</tr>
<tr>
<td><strong>Congenital</strong></td>
<td>The result of something that happens within the uterus or during delivery.</td>
</tr>
<tr>
<td><strong>Cranial</strong></td>
<td>Concerning the skull.</td>
</tr>
<tr>
<td><strong>Dandy-Walker Cyst</strong></td>
<td>A fluid filled sac, in the area of the fourth ventricle, causing a blockage.</td>
</tr>
<tr>
<td><strong>Distal</strong></td>
<td>The far end (often used to refer to the end of the catheter farthest from the brain).</td>
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<tr>
<td><strong>Fontanelle</strong></td>
<td>Two openings – “soft spots” in the bones of the skull. Located at the top of the head, one fills in around the 3rd/4th month and the other later, but before the age of two years.</td>
</tr>
<tr>
<td><strong>Hemorrhage</strong></td>
<td>Bleed</td>
</tr>
<tr>
<td><strong>Inguinal hernia</strong></td>
<td>A pouch in the muscle wall in the groin area.</td>
</tr>
<tr>
<td><strong>Intracranial</strong></td>
<td>Within the skull.</td>
</tr>
<tr>
<td><strong>Intraventricular hemorrhage</strong></td>
<td>Bleeding within the ventricles.</td>
</tr>
<tr>
<td>Term</td>
<td>Definition</td>
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<td>-------------------------------</td>
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<tr>
<td><strong>Ischemia</strong></td>
<td>A condition which an organ is cut off from its blood supply.</td>
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<tr>
<td><strong>Meninges</strong></td>
<td>A three layered membrane covering the brain and spinal cord.</td>
</tr>
<tr>
<td><strong>Meningitis</strong></td>
<td>An infection of the meninges.</td>
</tr>
<tr>
<td><strong>Non-communicating hydrocephalus</strong></td>
<td>Hydrocephalus caused by a blockage within the ventricles.</td>
</tr>
<tr>
<td><strong>Papilledema</strong></td>
<td>Swelling of the optic nerve (nerve which runs between the brain and the eye) at the point of entry to the eye. Usually caused by intracranial pressure.</td>
</tr>
<tr>
<td><strong>Parinaud’s syndrome</strong></td>
<td>Downcast eyes, eyes which do not move together or cross.</td>
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<tr>
<td><strong>Peritoneal cavity</strong></td>
<td>Abdominal cavity.</td>
</tr>
<tr>
<td><strong>Porencephaly</strong></td>
<td>An absence of a section of the brain which connects to the ventricles.</td>
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<tr>
<td><strong>Proximal</strong></td>
<td>Closest section (in this context usually refers to the section of the shunt closest to the head).</td>
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<tr>
<td><strong>Revision</strong></td>
<td>Surgery to repair/replace a malfunctioning shunt.</td>
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<tr>
<td><strong>Sunsetting eyes</strong></td>
<td>A condition in which the eyes seem to be downcast all the time. Too much white of the eye is visible above the iris.</td>
</tr>
<tr>
<td><strong>Suture lines</strong></td>
<td>The joins between the bones of the skull.</td>
</tr>
<tr>
<td><strong>Ventricular system</strong></td>
<td>The interconnecting system of four ventricles and the channels which link them – the Foramen of Munro and the Aqueduct of Sylvius.</td>
</tr>
<tr>
<td><strong>Ventriculitis</strong></td>
<td>An infection of the ventricular system.</td>
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</tbody>
</table>