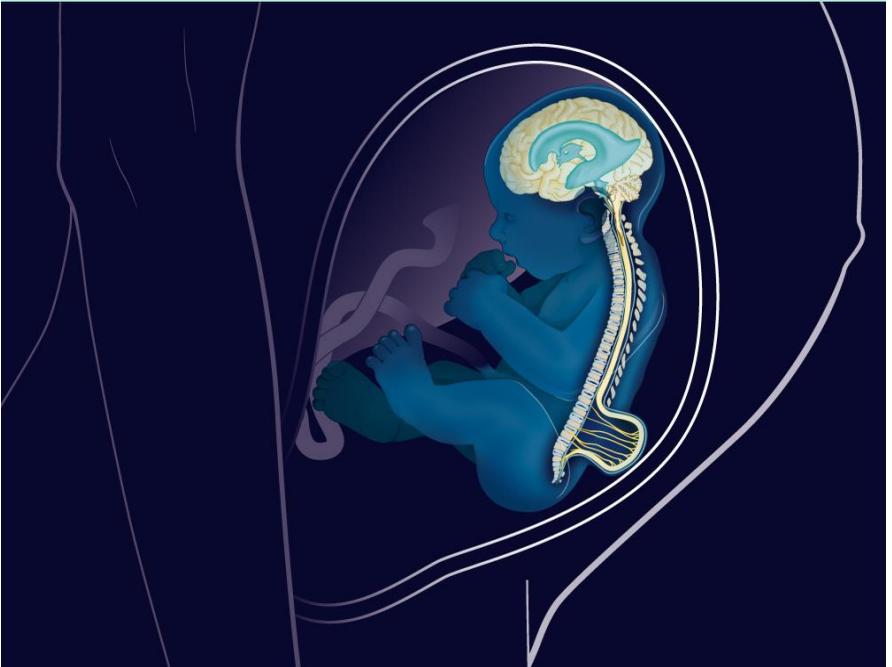


University College Hospital

Management of Fetal Spina Bifida Information for Pregnant Women & Families



Contents

1

1. Introduction	2
2. What is spina bifida?	2
3. What causes spina bifida?	4
4. What does it mean for my child?	4
4a. Motor, sensory and orthopaedic problems	5
4b. Brain changes	5
4c. Bladder and bowel problems	6
4d. Hormonal problems	6
4e. Psychosocial and sexual problems	7
5. Should I continue or terminate my pregnancy?	7
6. What if I decide to terminate the pregnancy?	7
7. What if I decide to continue the pregnancy?	8
8. What is fetal surgery?	9
9. How can fetal surgery help?	9
10. What are the risks of fetal surgery?	10
11. What will happen if I choose not to have surgery?	10
12. How should I prepare for fetal surgery?	10
13. Asking for consent	11
14. What happens during fetal surgery?	12
15. What should I expect after fetal surgery?	12
16. Contact details	13
17. Space for notes and questions	14

1 Introduction

You have been given this leaflet because your unborn baby (fetus) has been diagnosed with spina bifida. This news must have been stressful for you and your partner/family, and you will have to handle a lot of information on your options in a short time span. You have been given this leaflet to help you to remember everything that was discussed during your counselling sessions.

2 What is spina bifida?

Spina bifida arises when, early in pregnancy (before the sixth week), the spinal column and spinal cord are not properly formed. It occurs in around 4 in 10,000 pregnancies. There are different forms of spina bifida:

- **Spina bifida occulta** (Latin for “hidden spina bifida”). The spinal cord and nerve tissue may not have formed properly but they are covered by normal skin. This covering provides some protection to the nerve tissue. Surgery is sometimes necessary for spina bifida occulta but this is usually carried out when the baby is a little older. There is no role for surgery before birth in these cases.
- **Spina bifida aperta** (Latin for “open spina bifida”). The spinal cord and nerve tissue bulge through a hole or area of thin membrane on the baby’s back. This form of spina bifida is also called meningocele or myelomeningocele. Since in spina bifida aperta, the nerve tissue is exposed on the surface there is a risk of additional nerve damage and infection. In this type of spina bifida, surgery is carried out very soon after the baby is born to try to reduce the risk of these problems

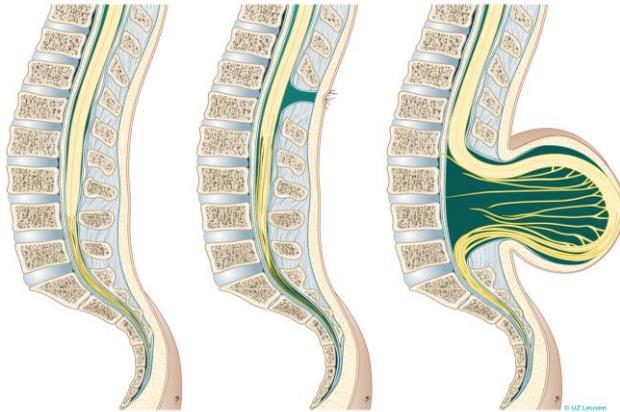


Diagram 1: Types of Spina Bifida

Left: normal situation / Middle: spina bifida occulta
 Right: spina bifida aperta

These different types are shown in diagram 1 above. Often with open spina bifida the hind brain descends through the opening in the skull where the spinal cord exits. This is called a Chiari II malformation. This may cause neurological problems after birth. It may also contribute to excessive accumulation of fluid in the brain ventricles. This is referred to as “ventriculomegaly”, a situation that may progress into hydrocephalus. This is shown in diagram 2.

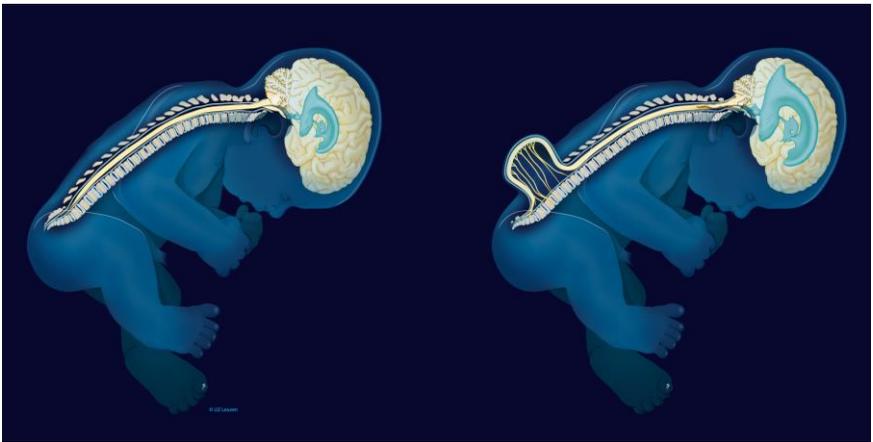


Diagram 2: Chiari II Malformation with Ventriculomegaly

3 What causes spina bifida?

The exact cause is unknown. Several factors may be responsible such as insufficient folate (a B vitamin) in the diet. There are also hereditary factors. Once spina bifida has occurred, there is a slightly increased risk of having a baby with spina bifida again in another pregnancy. Because of this we advise you to take a higher dose of folic acid (5 mg per day instead of the usual 0.4 mg per day) prior to conceiving a next pregnancy. Your doctor should refer you to a genetic specialist (geneticist) if there are several cases of spina bifida in your immediate family.

4 What does it mean for my child?

Every person with spina bifida is unique, meaning there is a large variety of possible effects, from mild to severe. Generally, a defect higher up on the back is associated with more serious consequences. Ultrasound of your baby can be used to determine the height of the defect in the spinal column before they are born. This will give doctors information about what problems the child will likely have. Typically these problems will include weakness in the feet or legs and problems with bladder and bowel control. However it is not possible to predict before birth exactly how severe these problems will be.

Sometimes other abnormalities are also found during the ultrasound scan. This combination increases the likelihood of an underlying genetic syndrome. A procedure to sample the amniotic fluid (amniocentesis) is recommended to exclude chromosomal problems (chromosomes are the carriers of genetic information inside each of our cells). If additional problems are confirmed, the outcome will depend on all abnormalities together. In the rest of this leaflet we will only discuss what we call an *isolated* spina bifida aperta or

isolated myelomeningocele. This means a baby with spina bifida aperta without other abnormalities.

4a Motor, sensory and orthopaedic problems

Normally, nerves from the spinal cord go to the muscles of the legs and feet to allow movement. In spina bifida, the spinal cord and nerves are often entirely or partially interrupted at the place of the lesion in the back. This can cause:

- curvature of the spine (also called scoliosis)
- muscle weakness in the legs
- impaired balance
- contracture (stiffness and tightening) of several joints
- club feet (also called talipes).

For children with a very low lesion, they are expected to be able to stand and learn to walk or have only mild weakness or curvature in the feet. For lesions at progressively higher parts of the spine, children may have difficulties with walking, or may not be able to walk, stand or even sit.

Because nerve pathways are interrupted in spina bifida, there is less or no sensation in the skin served by nerves below the site of the lesion in the back. This can lead to pressure ulcers in the skin, or it can cause cold feet and lower legs.

4b Brain changes

The changes in the hindbrain, referred to as the Chiari II or Arnold-Chiari malformation do not necessarily lead to problems for the child, and will not usually require treatment. However, in severe cases children may have difficulty with swallowing and breathing.

Additionally brain fluid may continue to accumulate inside the

ventricles, leading to ventriculomegaly. This occurs in about half to two thirds of babies with spina bifida. If untreated, hydrocephalus can cause damage to the brain due to increasing pressure.

When hydrocephalus occurs, surgeons often need to implant a shunt. This is a thin tube that drains the fluid from the brain into the abdominal cavity (tummy) of the child. The satisfactory working of the shunt will be assessed from time to time in the out patient clinic. Further surgery to a shunt may be required if there are problems such as infection or blockage. Such shunt complications can be responsible for long term disabilities. Hydrocephalus and complications of the drains may result in some learning and behavioural issues.

As many as two thirds of children with spina bifida have an IQ within the normal range. However, children with open spina bifida may have learning and behaviour issues. These include difficulties with concentration, attention and problem solving.

4c Bladder and bowel problems

Children with spina bifida often have problems controlling their bladder. This may cause urinary incontinence or inability to fully empty their bladder (referred to as urinary retention). This may lead to bladder and/or kidney infections. To prevent complications, medical staff may recommend that they empty their bladder using a small catheter through the urethra several times a day (called clean intermittent catheterisation). First the parents are taught this and later the children can learn to do it for themselves.

Children also often have bowel issues, resulting in either incontinence or constipation. This can be helped by bowel wash-outs (irrigation) or medication.

4d Hormonal problems

Children with spina bifida often have a shorter stature than usual. Early puberty is more common, especially in cases of hydrocephalus.

4e Psychosocial and sexual problems

When young adults with spina bifida are asked about whether they feel happy, their response usually is very similar to their peers. Men with spina bifida may have decreased fertility, but medication usually helps with erection difficulties. Women with spina bifida may have reduced sensation which affects their full enjoyment of sex.

5 Should I continue or terminate my pregnancy?

Many factors play a role when choosing what to do in your pregnancy. These include the anticipated severity of the condition and how you and your partner and family feel you can cope with caring for a child with this condition. In the UK there is no legal limit on when a termination for spina bifida can be performed. If you want support when taking this decision, your doctor can put you and your partner in touch with a counsellor or psychologist.

6 What if I decide to terminate the pregnancy?

If this is your choice then your labour and the delivery will usually be medically induced. This is done in hospital. Medical staff will discuss the entire process with you beforehand. After delivery you will have time to spend with your child, and the opportunity to take photographs if you wish. Staff can arrange a cremation or burial, but that is not

mandatory. Your doctor may suggest that you have additional testing (such as genetic or chromosome tests, or a postmortem of the baby) after delivery. This will depend on what exactly is seen on scans.

7 What if I decide to continue the pregnancy but not have fetal surgery?

If this is your choice then you will be supported throughout your pregnancy. During the last three months of pregnancy, your child will be monitored by ultrasound, for instance to measure the amount of fluid in the brain of the baby. If the fluid increases, sometimes medical staff will recommend delivering your baby a bit earlier. The delivery should be done in a centre where a spina bifida medical team is available, so that your child can be taken care of by a specialist team. Some women can have a vaginal delivery without this damaging the baby's spinal cord lesion.

During the first days of life, several specialists (including a paediatric neurologist, neurosurgeon, urologist or orthopedic surgeon) may examine your child. Medical staff may arrange a number of imaging tests, such as ultrasound or MRI (magnetic resonance imaging), in order to measure the effect of the spina bifida on the brain and the spinal cord. This information will help plan further management and predict the long-term outcome as accurately as possible. The team looking after your baby will include all kinds of specialists, such as a specialist nurse, paediatrician, social worker and paediatric physiotherapist.

An operation to close the defect is needed very soon after birth. The surgery is performed by a paediatric neurosurgeon, sometimes with a plastic surgeon. The surgeons will discuss the details of the operation with you beforehand. The operation usually takes less than two to

three hours. Thereafter, the baby will be regularly checked, in particular to assess whether they may need a shunt.

8 What is fetal surgery?

Fetal surgery is where surgeons perform an operation on a baby whilst it is still in the womb, hence before it is born.

In a baby with spina bifida, the defect is closed using a similar surgical technique to closure after birth. The womb is then closed and the baby continues to grow and develop in the womb until birth. In 2011, the results of an American study called the "MOMS trial" were published. The study investigated the effect of operating on babies with spina bifida whilst they were still in the womb as compared to operating on babies *after* birth.

Because of the benefits shown in that study, NHS England has commissioned two providers to provide this surgery University College Hospitals London and University Hospitals Leuven, Belgium.

Because it is a new treatment in the UK, information on your care during pregnancy and after birth will be collected and reviewed so that we can measure the quality of our care.

9 How can fetal surgery help?

In the MOMS study mentioned above, there were 77 children with spina bifida who were operated on in pregnancy, while they were still in the womb. Their outcomes were compared to 80 babies who had spina bifida surgery after birth.

Children who were operated before birth had:

- Less severe Chiari II malformation (hindbrain herniation)
- Less need for a shunt for hydrocephalus by the age of one year, though the effect was less when the ventricles were already enlarged before the operation.
- Better leg function at 30 months of age

On further follow-up those operated before birth seemed to have better bladder function, but that evidence is not yet conclusive. Follow-up research should make that clear.

10 What are the risks of fetal surgery?

There are risks of performing the spina bifida closure operation before birth, such as an increased risk of premature labor, premature rupture of membranes and decreased amniotic fluid. Premature delivery (birth before 37 weeks of gestation) can have its own complications that may offset some benefits of the fetal surgery.

After open fetal surgery, doctors recommend that women deliver their babies by Caesarean section, both in the same pregnancy and in future pregnancies. This is to prevent the scar in the womb opening during labour. Women are also asked to wait two years before conceiving again.

11 What will happen if I choose not to have fetal surgery?

Your care will not be negatively affected; your wishes will be respected and your care will continue as usual (see sections 6 and 7).

12 How should I prepare for fetal surgery?

If you are interested in fetal surgery for your baby, your doctor will discuss it with you and your partner.

Doctors will repeat the evaluation of your baby by ultrasound and MRI, before the decision can be made to proceed with surgery. This is to make sure that we know as much as possible about the condition that your baby has.

The fetal surgery operation should take place before 26 weeks of gestation. Fetal surgery is only considered when the lesion is open (this is the most common type of spina bifida), in a singleton pregnancy (i.e. not twins or triplets), and when we know that the chromosomes of the baby are normal.

The level of the lesion has to start at the thoracic or lumbar or upper sacral vertebrae, and there has to be hindbrain herniation (Chiari II malformation).

Surgery during pregnancy is not done in cases where one or more criteria are not met including:

- there are additional abnormalities with the baby (anatomical or chromosomal)
- there is an increased risk of premature birth (e.g. previous premature birth, shortened cervix)
- there is maternal obesity (BMI above 40)
- previous surgery on the womb (apart from caesarean section)
- there is a low-lying placenta.

If fetal surgery is not an option, surgery will be offered to your child after birth.

13 Asking for your consent

If you decide to go ahead with fetal surgery, by law we must ask for your permission and we will ask you to sign a consent form. This confirms that you agree to have the procedure

and understand what it involves. Staff will explain all the risks, benefits and alternatives before they ask you to sign a consent form. If you are unsure about any aspect of your proposed treatment, please don't hesitate to speak with a senior member of staff again.

Whether you decide to proceed with open fetal surgery or not, we will also ask you if we can collect anonymised information on our pregnancy so that we can audit our service for quality purposes.

14 What happens during fetal surgery?

You will be asked to come into hospital after fasting the night before. You will be shown to a room and give a gown to change into. When you are ready you will be taken into the operating theatre and the anaesthetist will offer to insert an epidural catheter (fine plastic tube) into your back for pain relief after the operation. You will then have a general anaesthetic (to put you to sleep). Once you are anaesthetised, a bikini line cut will be made in your abdomen (tummy). This is slightly wider than the size of the cut used for Caesarean section birth, and slightly higher up on the abdomen. The uterus (womb) is then opened to allow access to the baby. The spina bifida defect is examined and closed by the paediatric neurosurgeon. The uterus and then abdomen are both closed. You will then be woken up.

15 What should I expect after fetal surgery?

When you wake up from the operation you may feel a bit sore, but painkillers will be available for you. You will also have an epidural analgesia catheter, through which you can get pain relief. There will be a catheter in your bladder which

will be removed once you are able to walk around and pass urine in the toilet.

You will stay in hospital for a period of around one week, and when you go home you will be offered regular follow up and advised to keep relative rest.

16 Contact details

If you would like more information about fetal surgery for spina bifida please contact us on the following:

Email: spinabifida-fetalsurgery@nhs.net

17 Space for notes and questions

You may have many questions following this diagnosis, which can be difficult to remember during consultations. Please write them down so you can discuss them with your doctors.

The internet is a source of a wealth of information, but please be cautious as information found can be confusing and inaccurate. Please ask your doctor for suggested websites.

It may be useful to remember who you have been speaking to in the hospital. You can write those names down on this page.

Date of counselling: _____

Obstetrician: _____

Paediatrician: _____

Paediatric neurosurgeon: _____

Others: _____

Space for your notes:

This information leaflet is based on the Dutch leaflet “Spina bifida en foetale behandeling tijdens de zwangerschap” which was created with the help of members of the PROSPER consortium (Prospective Spina Bifida Registry of Outcome after Intrauterine Surgery), a national multidisciplinary group of experts involved in the pre- and post-natal care for children with spina bifida.

Images by Myrthe Boymansm, reproduced with permission from the Fetal Medicine Unit, UZ Leuven, Belgium.

This information leaflet has been reviewed by SHINE charity: Spina bifida • Hydrocephalus • Information • Networking • Equality www.shinecharity.org.uk

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We are committed to
delivering top-quality patient
care, excellent education
and world class research

Safety
Kindness
Teamwork
Improving

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