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Percutaneous fetal balloon valvuloplasty for pulmonary atresia with intact ventricular septum

Understanding NICE guidance – information for people considering the procedure, and for the public
Ordering information
You can download the following documents from www.nice.org.uk/IPG176
- this booklet
- the full guidance on this procedure.
For printed copies of the full guidance or information for the public, phone the NHS Response Line on 0870 1555 455 and quote:
- N1043 (full guidance)
- N1044 (information for the public).
About this information

The National Institute for Health and Clinical Excellence (NICE) is the independent organisation responsible for providing national guidance on the promotion of good health and the prevention and treatment of ill health. One of NICE’s roles is to produce guidance (recommendations) on whether interventional procedures are safe enough and work well enough to be used routinely in the NHS in England, Wales and Scotland.

This information describes the guidance that NICE has issued on a procedure called percutaneous fetal balloon valvuloplasty for pulmonary atresia with intact ventricular septum. It is not a complete description of what is involved in the procedure – the healthcare team should describe it in detail.

NICE has looked at whether percutaneous fetal balloon valvuloplasty is safe enough and works well enough for it to be used routinely for the treatment of pulmonary atresia with intact ventricular septum.

To produce this guidance, NICE has:

● looked at the results of studies on the safety of percutaneous fetal balloon valvuloplasty and how well it works
● asked experts for their opinions
● asked the views of the organisations that speak for the healthcare professionals and the parents and carers who will be affected by this guidance.

This guidance is part of NICE’s work on ‘interventional procedures’ (see ‘Further information’ on page 10).
About the procedure

Pulmonary atresia is a heart defect that may be present in a baby when it is born and when it is still in the womb (when it is called a fetus). Babies in the womb have slightly different blood systems compared with babies that are a few days old. Before birth a baby has a special blood vessel in the heart (called the ductus arteriosus) and a connection between the two upper chambers of the heart (called the foramen ovale). These allow the blood to bypass the lungs because, in the womb, the baby gets oxygen from its mother's blood and doesn’t use its lungs. Both the ductus arteriosus and the foramen ovale usually close up when the baby is born, or soon after.

In pulmonary atresia the heart’s pulmonary valve is completely closed, which prevents the flow of blood from the heart to the lungs to ‘collect’ essential oxygen. Instead the blood passes from the right side of the heart to the left side of the heart through the foramen ovale or through any defects (if there are any) in the septum, which is the partition between the two lower chambers of the heart or ventricles. Some blood will then flow to the body and some will pass to the lungs via the ductus arteriosus.

After birth, however, blood can only get to the lungs if the foramen ovale, any other defects and the ductus arteriosus remain open. Babies who have pulmonary atresia with intact ventricular septum (shortened to PAIVS) do not have any defects in the septum. If the foramen ovale and the ductus arteriosus close up soon after birth, these babies become short of oxygen.

When babies with PAIVS are born parts of their hearts are not properly developed, and about 3% (which is 3 in every 100) die before they are born from hydrops, a condition in which too much fluid builds up in their tissues. Rarely babies have severe PAIVS and these babies are very likely to be very sick or to die soon after they are born.
Many babies with PAIVS survive until they are born and can then be treated. About half of the children who have PAIVS have a heart where both ventricles work properly. About 85% of babies who are born with PAIVS live until they are 5 years old. Because some of these babies and children need several stages of complicated operations where the chest is opened up to get at the heart (open-heart surgery) some parents may choose to have an abortion.

Babies who are born with PAIVS are treated in stages. A procedure in which a small balloon is inserted via the blood vessels to open up the baby’s pulmonary valve is usually done first to help the baby’s heart develop more normally. This is known as postnatal balloon valvuloplasty. These babies usually have more similar procedures and the valve is usually replaced later. If these procedures are not successful the baby may need open-heart surgery. There is a risk that the baby will die during or after these procedures.

Fetal balloon valvuloplasty is a procedure to open up the closed pulmonary valve of the baby’s heart while it is still in the womb. The procedure is intended to help the heart to develop more normally before the baby is born and to prevent hydrops from which the baby may die. This may increase the chance that operations on the baby’s heart after it is born will be successful.

This procedure may be considered for babies when there is a high risk that their heart condition will get worse before they are born and that they may be very sick or die soon after birth.

Fetal aortic balloon valvuloplasty is done at 21 to 32 weeks into the pregnancy. The mother is given a local anaesthetic to numb the area and a needle is inserted through the mother’s skin into the womb using ultrasound images to guide the needle. A painkiller is injected into the baby. The needle is then inserted through the unborn baby’s chest wall into its heart. A wire is put in through the needle across the pulmonary valve. A thin tube (catheter) with a special balloon is
inserted and the balloon is inflated to expand the closed valve so blood can flow through it. The tube and the needle are then removed.

The procedure can only be done successfully if the baby is in a suitable position inside the womb.

How well the procedure works
What the studies said
Not many studies have been published about this procedure. The reported studies cover less than 10 cases altogether, where babies most likely to benefit from the procedure were very carefully chosen. The largest study looked at five babies after fetal pulmonary valvuloplasty. The valve was opened up successfully in three babies and their blood flow was improved. All three babies survived to have more operations after they were born – one was 2, one was 3 and the other was 4½ years when the report was written. The two babies for whom the procedure was not successful were both born alive but died soon after they were born, although one of them had another operation after it was born.

It is difficult to tell how well the procedure works because so few babies have had it and because the problems of the babies before they had the procedure varied.

None of the studies mentioned how the mothers were affected.

What the experts said
The experts noted that there was not much evidence about the procedure. They said that it is difficult to judge how well the procedure works based only on how long the babies live, when the problem is rare and babies are carefully chosen to have the procedure.
Risks and possible problems with the procedure

What the studies said
Not much information has been published about the safety of the procedure. In one study two babies had increase in fluid in the sac around the heart (called pericardial effusions) that got better on its own. In a study of five babies one had pericardial effusions for a brief time but this got better on its own.

None of the studies mentioned whether the mothers had any problems.

What the experts said
The experts said that the possible problems that might be caused by the procedure included the baby dying, bleeding, slow heart beat, pericardial effusion and rupture of the balloon. They also mentioned that there was a risk that labour might start prematurely and there might be health problems for the mother because of the use of anaesthesia.

What has NICE decided?
NICE has decided that, if a doctor wants to carry out percutaneous fetal balloon valvuloplasty, he or she should make sure that the mother understands what is involved and that there are still uncertainties over the safety of the procedure and how well it works. There should be special arrangements in place so that the mother only agrees (consents) to the procedure after this discussion has taken place. Clear written information should be given to the parents as well as counselling and support both before and after the procedure.

NICE has said that the procedure should only be done in hospitals that specialise in doing procedures on babies while they are still in the womb. It should only be done where there is a team of specialists
including a consultant in fetal medicine, a children’s heart specialist, a specialist in dealing with newborn babies, a specialist midwife and a children’s heart surgeon.

A central store of information is being started by the Association for European Paediatric Cardiology and doctors are encouraged to enter details of all of their cases so that the safety of the procedure and how well it works can be checked over time.

NICE has said that more reports about how to choose which unborn babies are most likely to benefit from the procedure would be useful, and NICE may look at the procedure again when more information is published.

**What the decision means for you**

Your doctor may have offered you percutaneous fetal balloon valvuloplasty. NICE has considered this procedure because it is relatively new. NICE has decided that there are uncertainties about the benefits and risks of percutaneous fetal balloon valvuloplasty which you need to understand before you agree to it. Your doctor should discuss the benefits and risks with you. Some of these may be described above.

Because more information is needed about percutaneous fetal balloon valvuloplasty NICE is encouraging the collection of some details about every mother who has the procedure in England and Wales. These details will be held confidentially and will not include parents’ names. The information will be used only to see how safe the procedure is and how well it works. If you decide to have percutaneous fetal balloon valvuloplasty, you will be asked to agree to your details being entered into an electronic database for this purpose. A clinician looking after you will fully explain the purpose of collecting the data and what details will be held.
Further information

You have the right to be fully informed and to share in decision-making about the treatment you receive. You may want to discuss this guidance with the doctors and nurses looking after you.

The NICE website (www.nice.org.uk) has further information about NICE, the Interventional Procedures Programme and the full guidance on percutaneous fetal balloon valvuloplasty that has been issued to the NHS. The evidence that NICE considered in developing this guidance is also available from the NICE website.

NICE has also issued guidance on: balloon valvuloplasty for aortic valve stenosis in adults and children (www.nice.org.uk/IPG078), balloon dilatation of pulmonary valve stenosis (www.nice.org.uk/IPG067), balloon angioplasty of pulmonary vein stenosis in infants (www.nice.org.uk/IPG075), balloon dilatation with or without stenting for pulmonary artery or non-valvar right ventricular outflow tract obstruction in children (www.nice.org.uk/IPG076), balloon dilatation of systemic to pulmonary arterial shunts in children (www.nice.org.uk/IPG077), radiofrequency valvotomy for pulmonary atresia (www.nice.org.uk/IPG095) and percutaneous fetal balloon valvuloplasty for aortic stenosis (www.nice.org.uk/IPG175).

If you have access to the internet, you can find more information on congenital heart disease on the NHS Direct website (www.nhsdirect.nhs.uk).

You can also phone NHS Direct on 0845 46 47.