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Percutaneous fetal balloon valvuloplasty for aortic stenosis

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/IPG175

- 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:

- N1041 (full guidance)
- N1042 (information for the public).

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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 aortic stenosis. 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 aortic stenosis.

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

Aortic valve stenosis 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.

Stenosis means narrowing and in aortic valve stenosis the aortic valve in the heart has become narrow. Normally this valve lets blood flow out of the heart into the main blood vessels of the body and stops it from flowing backwards. When the aortic valve becomes narrow blood can't flow out of the heart easily, which puts a strain on the heart. In severe aortic stenosis blood is unable to pass into the left ventricle which can lead to hypoplastic left heart syndrome or HLHS for short. In HLHS all of the structures on the left side of the heart may be very malformed or severely underdeveloped. This means they are unable to work properly and support biventricular circulation.

The blockage of the blood leads to increased pressure in the left side of the heart. If the foramen ovale is open blood flow passes from left to right; however, if the foramen ovale closes before birth the resulting pressure produces scar-like tissue in the heart. This condition is known as aortic stenosis with a restrictive interatrial communication.

These defects may not affect the baby very much or in rare cases they may be severe and make it very likely that the baby will be very ill or will die soon after it is born. Many babies with severe aortic stenosis survive until they are born but about one in ten die in the womb. Because of their illness biventricular heart repair is not a suitable treatment for most babies with severe aortic stenosis. About half of babies born with severe aortic stenosis die during their first year of life despite having operations to treat their problem. Because the chance that the babies will not survive is so high, parents of babies with severe aortic stenosis may choose to have an abortion.

For babies who are born with aortic valve stenosis but whose heart is not too severely affected, the valve is usually repaired by passing a small balloon into the heart through the blood vessels. This helps the heart to grow more normally. Further operations are often needed to repair the valve again and then to replace it.

Babies who are born with HLHS may need up to three operations taking 3 years or longer to make their heart work more normally. These operations involve complicated surgery in which their chest is opened to get at the heart (open-heart surgery). There is a risk that the baby will die during or after these operations.

Fetal aortic balloon valvuloplasty is a procedure to widen the narrow stenotic valve of the the baby's heart while it is still in the womb. The aim of the procedure is to help the heart to develop more normally before the baby is born. 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 chest wall of the baby into its heart. A wire is inserted through the needle across the aortic valve. A thin tube (catheter) with a special balloon is then inserted and the balloon is inflated to expand the narrowed valve so blood can flow through it more easily. 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. In a group of 20 babies who had the procedure, the valve was expanded successfully in 14 of them. In these 14 babies their heart valves and the main blood vessel leading away from the heart (aorta) grew better compared with 10 babies who did not have the procedure or for whom it was not successful. Among the 14 babies who had successful procedures, 3 were born with a biventricular heart, 6 were born with HLHS, 2 died in the womb and another 3 were not yet born when the report was written.

In another group of 12 babies with severe blockages of the aortic valve who were given the procedure, the valve was expanded successfully in 7.

Not much is known about how well the mothers do after the procedure. In one study the mothers stayed in hospital for an average of 2 days but this ranged from 1 to 7 days.

What the experts said

The experts noted that not much is known about this procedure. They stated that it is difficult to judge how well the procedure works based only on how long the babies live, when the condition is rare and babies are carefully chosen to have the procedure.

Risks and possible problems with the procedure

What the studies said

In a study of 20 babies, three died because of the procedure, two within a day of the procedure, one with severe accumulation of fluid in the tissues (hydrops) and one with stress. The third one died 3 days after the procedure because of a very slow heart beat

(bradycardia). In this study, 15 of the 20 babies had slow heartbeats and 11 of these needed drug treatment for this.

In a second study of 12 babies, 4 died within 24 hours of the procedure, 2 from persistent slow heartbeat (bradycardia), 1 from bleeding, and 1 when the valve was cut open after an emergency delivery. In both studies there were also problems with the balloon rupturing and the baby's heartbeat becoming slow during the operation.

The main problems in the mothers were to do with needing to have an emergency caesarean operation after the fetal valvuloplasty procedure. This operation was needed in 3 out of 12 babies in one study. The caesarean was done because the baby had a slow heartbeat in 2 cases and because of inflammation of the membranes that surround the baby in the womb caused by an infection in the third.

What the experts said

The experts said that, as well as the possibility that the baby would die, bleeding, slow heart beat, increase in fluid in the sac around the heart (known as pericardial effusion) and the balloon rupturing were all possible problems. 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 for aortic stenosis, 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 for aortic stenosis. 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 parent 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 for aortic stenosis 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 pulmonary atresia with intact ventricular septum (www.nice.org.uk/IPG176).

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.

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