NICE guidance

Open prenatal repair for open neural tube defects in the fetus

Interventional procedures guidance
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www.nice.org.uk/guidance/ipg668

1 Recommendations

1.1 Evidence on the efficacy of open prenatal repair of open neural tube defects in the fetus is adequate in quantity and quality. However, evidence on its safety shows serious but well recognised safety concerns for the mother and fetus. Therefore, this procedure should only be used with special arrangements for clinical governance, consent, and audit or research. Find out what special arrangements mean on the NICE interventional procedures guidance page.

1.2 Clinicians wishing to do open prenatal repair of open neural tube defects in the fetus should:

- Inform the clinical governance leads in their NHS trusts.
- Give parents clear written information to support shared decision making, including NICE's information for the public.

- Ensure that parents understand the procedure's safety and efficacy, as well as any uncertainties about these.
- Audit and review clinical outcomes of everyone having the procedure. NICE has identified relevant audit criteria and has developed an audit tool (which is for use at local discretion).

1.3 The procedure is technically challenging and should only be done in specialised centres, and only by clinicians and teams with specific training and experience in open prenatal repair.

1.4 Patient selection should only be done by a multidisciplinary team, which should include a consultant in fetal medicine, an obstetric surgeon, a paediatric neurosurgeon, a radiologist with experience in fetal imaging and an anaesthetist.

1.5 Further research should report details of risks to the mother (including her subsequent pregnancies), risks to the fetus (including the need for further surgery) and long-term disability after birth.

2 The condition, current treatments and procedure

The condition

2.1 Neural tube defects happen because the neural tube does not fuse during early embryonic development. Open neural tube defects are those in which the affected region of the neural tube is exposed on the body's surface. The most common neural tube defect is spina bifida, where the defect is in the spine. Myelomeningocele (open spina bifida) is the most severe type of spina bifida, in which the baby's spinal canal remains open along several vertebrae in the back. The spinal cord and protective membranes around it push out and form a sac, which is exposed on the baby's back. Children born with myelomeningocele may experience motor neurological deficits including muscle weakness and paralysis of the lower limbs, sensory deficit, bowel, bladder and sexual dysfunctions, and learning difficulties. The condition can be associated with Chiari II

malformation (hindbrain herniation) and hydrocephalus.

Current treatments

2.2 Conventional treatment for myelomeningocele (open spina bifida) is immediate surgical repair of the defect within days of birth to prevent further damage to nervous tissue and reduce the risk of central nervous system infection. The immediate management may also include ventricular-peritoneal shunt placement to relieve hydrocephalus. The condition can also be treated prenatally with the aim of decreasing morbidity in the child.

The procedure

2.3 Open prenatal repair for open neural tube defects is typically done before 26 weeks of pregnancy. Using general anaesthesia, a low transverse laparotomy incision is done and the gravid uterus is exposed and exteriorised. The fetus and placenta are visualised by ultrasound, and the fetus is manually positioned to allow a uterine incision (hysterotomy) over the centre of the myelomeningocele sac. The hysterotomy location is either anterior, fundal or posterior depending on the location of the placenta. The hysterotomy is made large enough to allow the neural tissue in the meningomyelocele to be dissected from surrounding tissue so that it can drop into the spinal canal. The defect is then closed. If there is insufficient dura or skin for closure, occasionally a biocellulose and dermal regeneration patch substitute may be used for repair. The uterine incision is closed and a sodium lactate solution with antibiotics is added to the uterus until the amniotic fluid index is normal. The maternal abdominal wound is then closed.

2.4 A number of variations to the procedure have been described and the technique is still evolving.

3 Committee considerations

The evidence

3.1 NICE did a rapid review of the published literature on the efficacy and safety of this procedure. This comprised a comprehensive literature search and detailed review of the evidence from 10 sources, which was discussed by the committee. The evidence included 5 systematic reviews and meta-analysis, 1 randomised controlled trial, 3 case series and 1 case report. It is presented in table 2 of the interventional procedures overview. Other relevant literature is in the appendix of the overview.

3.2 The specialist advisers and the committee considered the key efficacy outcomes in the baby to be: motor function, hind brain herniation, hydrocephalus, bowel and bladder function, and need for further surgery.

3.3 The specialist advisers and the committee considered the key safety outcomes in the baby to be: fetal mortality, perinatal death, premature birth, premature rupture of membranes, cerebrospinal fluid leakage and the potential for late spinal cord complications including tethered spinal cord and syringomyelia. Key safety outcomes for the mother are: operative morbidity, incisional hernia, uterine dehiscence or rupture in the current or subsequent pregnancy, and morbidly adherent placenta in subsequent pregnancies.

3.4 Patient commentary was sought but none was received.

Committee comments

3.5 The committee noted the need to identify the risks and benefits for both the fetus and mother (including her subsequent pregnancies) including long-term outcomes, and that these need to be discussed during parental counselling by the multidisciplinary team.

3.6 The committee noted that some of the data it considered was for operations done at a gestational age above 26 weeks.

3.7 This guidance requires that clinicians doing the procedure make special arrangements for audit. NICE has identified relevant audit criteria and has developed an audit tool (which is for use at local discretion).

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Endorsing organisation

This guidance has been endorsed by Healthcare Improvement Scotland.

Accreditation