Combined bony and soft tissue reconstruction for hip joint stabilisation in proximal focal femoral deficiency (PFFD)

Interventional procedures guidance
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1 Guidance

1.1 Current evidence on the safety and efficacy of combined bony and soft tissue reconstruction for hip joint stabilisation in proximal focal femoral deficiency (PFFD) is inadequate in quantity, quality and consistency. Therefore this procedure should only be used with special arrangements for clinical governance, consent and audit or research.

1.2 Clinicians wishing to undertake combined bony and soft tissue reconstruction for hip joint stabilisation in PFFD should take the following actions.

- Inform the clinical governance leads in their Trusts.
• Ensure that parents or carers understand the uncertainty about the procedure's safety and efficacy. They should understand that multiple procedures may be needed and that the procedure may not result in a fully functioning limb. Parents or carers should be provided with clear written information. In addition, the use of NICE's information for patients ('Understanding NICE guidance') is recommended.

• Audit and review clinical outcomes of all patients having combined bony and soft tissue reconstruction for hip joint stabilisation in PFFD (see section 3.1).

1.3 The procedure should only be carried out in units that specialise in limb reconstruction, by surgeons with specialist knowledge of neonatal hip dysplasias and expertise in limb lengthening procedures.

2 The procedure

2.1 Indications and current treatments

2.1.1 Proximal focal femoral deficiency is a congenital syndrome typically characterised by poor hip joint development and femoral shortening. The severity of the syndrome is variable. In severe cases, there may be no hip joint and the femur may be very short. In addition, PFFD may be associated with other lower limb abnormalities, such as an abnormal knee joint, lower limb malrotation, inadequacy of the proximal musculature and limb length discrepancy.

2.1.2 Treatment options depend on the extent of the PFFD. In patients with severe forms of PFFD, it may not be possible to produce a leg that is functional and of the correct length, so partial limb amputation and fitting of a prosthesis may be the preferred management. In patients with relatively mild PFFD, an attempt can be made to correct the abnormalities of the hip joint and the upper femur.

2.2 Outline of the procedure

2.2.1 Combined bony and soft tissue reconstruction for hip joint stabilisation in PFFD is carried out with the patient under general anaesthesia. There are
several variations on the procedure. Hip stabilisation involves a long incision on the outer side of the thigh. With the soft tissues retracted or released, the upper femur deformity is corrected by bone division and fixation. If needed, the pelvic bone may also be divided and moved to help reconstruct the hip joint. After surgery, the joint may need to be immobilised in a plaster cast. If the hip joint cannot be salvaged, the upper femur may be stabilised against the pelvis using a pelvic support osteotomy, and this may be combined with leg lengthening procedures.

2.2.2 Several additional procedures may be required to achieve reconstruction or to enable prosthetic attachment, either at the same time or afterwards as separate procedures. These may include leg lengthening, epiphysiodesis of the normal (opposite) femur, knee reconstruction, Van Nes rotationplasty, and 'above-the-knee' amputation.

Sections 2.3 and 2.4 describe efficacy and safety outcomes which were available in the published literature and which the Committee considered as part of the evidence about this procedure. For more detailed information on the evidence, see the overview.

2.3 Efficacy

2.3.1 A case series of 14 patients reported that 64% (9/14) of patients had a good clinical outcome after hip stabilisation, leg lengthening and external fixation (based on a composite measure of gait, range of movement, degree of dislocation and residual shortening), at a mean follow-up of 17 years. At final follow-up, the mean difference in patients' limb length was 11.6 cm (range 1–20 cm) (duration of follow-up not stated). Angular deformity was reported in 21% (3/14) of patients (mean follow-up 17 years).

2.3.2 A case report of 3 patients who had plaster casts for 3 months then valgus osteotomy described successful reorientation and stabilisation of the hip and straightening of the femur. Femoral lengthening was undertaken in 1 patient and planned in 2 others at a follow-up of 2.3–8 years. The case series of 14 patients reported that 43% (6/14) needed more than one lengthening procedure.
2.3.3 The Specialist Advisers considered key efficacy outcomes to be overall limb function and a reduced need for repeat procedures. The Specialist Advisers also stated that for some patients, the result may not be as good as would have been achieved by amputation and fitting of a prosthesis.

2.4 Safety

2.4.1 The case series of 14 patients reported osteitis in 43% (6/14) of patients, fracture (not otherwise described) in 7% (1/14) of patients, and pseudoarthritis in 7% (1/14) of patients (mean follow-up 17 years).

2.4.2 The Specialist Advisers stated that adverse events (reported in the literature or anecdotally) include significant hip and knee stiffness as a result of excessive lengthening, hip dislocation and recurrent deformity. The Specialist Advisers also considered theoretical adverse events to include avascular necrosis, bone non-union, infection, nerve or vascular injury, poor limb function, recurrence of contractures and wound dehiscence.

3 Further information

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

3.2 NICE has published interventional procedures guidance on intramedullary distraction for lower limb lengthening.

Information for patients

NICE has produced information on this procedure for patients and carers ('Understanding NICE guidance'). It explains the nature of the procedure and the guidance issued by NICE, and has been written with patient consent in mind.
4 About this guidance

NICE interventional procedure guidance makes recommendations on the safety and efficacy of the procedure. It does not cover whether or not the NHS should fund a procedure. Funding decisions are taken by local NHS bodies after considering the clinical effectiveness of the procedure and whether it represents value for money for the NHS. It is for healthcare professionals and people using the NHS in England, Wales, Scotland and Northern Ireland, and is endorsed by Healthcare Improvement Scotland for implementation by NHSScotland.

This guidance was developed using the NICE interventional procedure guidance process.

We have produced a summary of this guidance for patients and carers. Tools to help you put the guidance into practice and information about the evidence it is based on are also available.

Changes since publication

7 January 2012: minor maintenance.

Your responsibility

This guidance represents the views of NICE and was arrived at after careful consideration of the available evidence. Healthcare professionals are expected to take it fully into account when exercising their clinical judgement. This guidance does not, however, override the individual responsibility of healthcare professionals to make appropriate decisions in the circumstances of the individual patient, in consultation with the patient and/or guardian or carer.

Implementation of this guidance is the responsibility of local commissioners and/or providers. Commissioners and providers are reminded that it is their responsibility to implement the guidance, in their local context, in light of their duties to avoid unlawful discrimination and to have regard to promoting equality of opportunity. Nothing in this guidance should be interpreted in a way which would be inconsistent with compliance with those duties.

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Accreditation

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