NATIONAL INSTITUTE FOR HEALTH AND CLINICAL EXCELLENCE

INTERVENTIONAL PROCEDURES PROGRAMME

Interventional procedure overview of combined bony and soft tissue reconstruction for hip joint stabilisation in proximal focal femoral deficiency (PFFD)

Children are sometimes born with defects of the hip joint and upper thigh bone called proximal femoral focal deficiency, often shortened to PFFD. Surgical treatment, aiming to produce as functional a leg as possible, may be appropriate. This may involve a corrective operation on the hip joint (sometimes referred to as the 'superhip' procedure). The initial operation may be combined with other surgery to lengthen the leg and treat other problems such as poor knee function at the same time or at a later date.

Introduction

The National Institute for Health and Clinical Excellence (NICE) has prepared this overview to help members of the Interventional Procedures Advisory Committee (IPAC) make recommendations about the safety and efficacy of an interventional procedure. It is based on a rapid review of the medical literature and specialist opinion. It should not be regarded as a definitive assessment of the procedure.

Date prepared

This overview was prepared in October 2008.

Procedure name

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

Specialty societies

- British Orthopaedic Association
- British Limb Reconstruction Society
- British Society for Children's Orthopaedic Surgery.

Description

Indications and current treatment

Proximal focal femoral deficiency (PFFD) involves poor hip joint development with femoral shortening and, in severe cases, failure of formation of the upper femur. The severity of the syndrome ranges from mild hip abnormality to a very short thigh bone and absent hip joint. In addition, PFFD may be associated with other lower limb abnormalities, such as abnormal knee joint, malrotation, inadequacy of the proximal musculature and limb length discrepancy.

Current treatment depends on the extent of the PFFD. In patients with relatively mild PFFD, an attempt can be made to correct the hip joint and upper femur abnormalities (this is sometimes called the ‘superhip’ procedure). Subsequently, the leg may be lengthened, usually using an external frame. If the hip joint cannot be salvaged, the upper femur may be stabilised against the pelvis using a pelvic support osteotomy with an external frame, and this may be combined with leg lengthening. The knee joint in PFFD may be stiff or unstable and this may need surgical correction.

In more severe forms of PFFD, it may not be possible to produce a leg which is functional and of the correct length. Partial limb amputation and fitting of a prosthesis may be the preferred management. Fusions or rotationplasty may be undertaken to optimise fitting of the artificial limb. Occasionally, if a child is predicted to be very tall, a decision may be made to limit growth of the opposite leg by epiphysiodesis.

What the procedure involves

There are a number of variations on the procedure used to bring about hip joint stability, but they usually entail one of the following two elements.

Hip reconstruction (sometimes referred to as the ‘superhip’ procedure)

A long incision is made on the outer side of the thigh. The outer muscles and tendons are moved aside and contracted soft tissues released as necessary. The upper femur deformity is corrected by bone division and fixation, which may require removal of the bent bone. If the femur is not continuous with the hip joint, the un-united area may be removed and the freshened bone ends fixed together to restore the integrity of the thigh bone. If the pelvic side of the hip joint is also poorly formed, the pelvic bone may be divided and moved to correct this problem. After the surgery the joint may need to be immobilised in a plaster cast.

Pelvic support osteotomy

An external scaffold (usually an Ilizarov frame) is attached to the outer femur. The femur is divided so the upper part can be placed beneath the pelvic bone to support the leg. Additional bone divisions may be needed in the pelvis and hip. This operation tends to shorten the already short femur further, and is often combined with a leg lengthening procedure.

A surgeon may undertake several additional procedures to assist reconstruction or to enable prosthetic attachment at the same time as the above procedures or afterwards as separate procedures. This may include leg lengthening, epiphysiodesis of the normal (opposite) femur, knee reconstruction, Van Nes rotationplasty, and 'above knee' amputation.

OPCS code

TBC

List of studies included in the overview

This overview is based on approximately 17 patients from one case series[1] and one case report[2].

Efficacy

One case series of 14 patients reported that 64% (9/14) of patients had a good clinical outcome (based on a composite measure of gait, range of movement, degree of dislocation and residual shortening) at a mean follow-up of 17 years[1]. At final follow-up, the mean residual shortening was 11.6 cm (range 1–20 cm). In this case series of 14 patients, angular deformity was reported in 21% (3/14) of patients and more than one lengthening procedure was required in 43% (6/14) of patients.

A case report of three patients described successful re-orientation and stabilisation of the hip and straightening of the femur, with femoral lengthening undertaken in one patient and planned in two others at a follow-up of 2.3–8 years[2].

Safety

In the case series of 14 patients fracture (not otherwise described) occurred in 7% (1/14), pseudoarthritis in 7% (1/14), and osteitis in 43% (6/14). Three out of 14 patients (21%) reported no complications[1].

Literature review

Rapid review of literature

The medical literature was searched to identify studies and reviews relevant to combined bony and soft tissue reconstruction for hip joint stabilisation in proximal focal femoral deficiency (PFFD). Searches were conducted of the following databases, covering the period from their commencement to 2 October 2008: MEDLINE, PREMEDLINE, EMBASE, Cochrane Library and other databases. Trial registries and the Internet were also searched. No language restriction was applied to the searches (see appendix C for details of search strategy).

The following selection criteria (table 1) were applied to the abstracts identified by the literature search. Where selection criteria could not be determined from the abstracts the full paper was retrieved.

Table 1 Inclusion criteria for identification of relevant studies

Characteristic	Criteria
Publication type	Clinical studies were included. Emphasis was placed on identifying good quality studies. Abstracts were excluded where no clinical outcomes were reported, or where the paper was a review, editorial, or a laboratory or animal study. Conference abstracts were also excluded because of the difficulty of appraising study methodology, unless they reported specific adverse events that were not available in the published literature.
Patient	Patients with limb deficiency in congenital proximal focal femoral deficiency.
Intervention/test	Combined bony and soft tissue reconstruction for hip joint stabilisation
Outcome	Articles were retrieved if the abstract contained information relevant to the safety and/or efficacy.
Language	Non-English-language articles were excluded unless they were thought to add substantively to the English-language evidence base.

Existing assessments of this procedure

There were no published assessments from other organisations identified at the time of the literature search.

Related NICE guidance

Below is a list of NICE guidance related to this procedure. Appendix B gives details of the recommendations made in each piece of guidance listed.

Interventional procedures

- Intramedullary distraction for lower limb lengthening. NICE interventional procedures guidance 197 (2006). Available from www.nice.org.uk/IPG197

Technology appraisals

- No relevant guidance

Clinical guidelines

- No relevant guidance

Public health guidance

- No relevant guidance

Table 2 Summary of key efficacy and safety findings on combined bony and soft tissue reconstruction for hip joint stabilisation in proximal focal femoral deficiency (PFFD)

Abbreviations used: PFFD, proximal femoral focal deficiency; ROM, range of movement

Study details	Key efficacy findings	Key safety findings	Comments
Fernandez-Palazzi F(2002)[1] Study type: **Case series** Country: Venezuela n = **22 (n = 14 combined procedure)** Study period: 1981 to 1990 Study population: PFFD, with the tip of the foot at medial tibial level on the contralateral limb. Age: 8.4 years (mean) (range 4–16 years) Sex: 50% male. Fixsen–Lloyd-Robert classification I (stable hip) = 59%, II = 36%, III = 14% Inclusion criteria: Not reported Technique: Hip stabilisation and subsequent leg lengthening and external fixation, with the Wagner, orthofix, or Lazo-Canadell, device. Distraction at a rate of 0.5 mm per day. Follow-up: **mean 17 years (for whole cohort)** Conflict of interest: Not reported.	**Subjective treatment outcome** Overall, 64% (9/14) of patients had a good outcome at final follow up, and 36% (5/14) were classified as having a bad outcome. In terms of subgroups based on PFFD radiological classification at baseline, of the 7 patients with Fixsen–Lloyd-Robert's grade I deficiency, 6 had a good outcome and 1 had a bad outcome. In 5 grade II patients, 2 had a good outcome and 1 had a bad outcome. In 2 grade III patients, 1 had a good outcome and 1 a bad outcome. **Limb deficiency** At final follow-up, the mean residual shortening was 11.6 cm (range 1–20 cm). **Repeat Procedures** 43% (6/14) of patients required more than one lengthening procedure. In 4 patients this was because of intolerance (requiring the treatment to be stopped to allow consolidation), and in 2 patients this was because of loosening or infection of pins. **Angular deformity** Angular deformity occurred in 21% (3/14) of patients.	~~**Complications**~~ ~~43% (6/14) of patients required more than one lengthening procedure. In 4 patients this was because of intolerance (requiring the treatment to be stopped to allow consolidation), and in 2 patients this was because of loosening or infection of pins.~~ **Event** — **Rate** Osteitis — 43% (6/14) Penetrant infection — 0% ~~Angular deformity~~ — ~~21% (3/14)~~ Pseudoarthritis — 7% (1/14) Fracture — 7% (1/14) No complication — 21% (3/14) Stiffness of the knee was reported in 43% (6/14) of patients following lengthening with complete extension and reduced flexion ROM.	This case series describes a mixed cohort of patients treated by hip stabilisation alone (for future orthosis), leg lengthening after stabilisation, and conservative management, depending on the severity of PFFD (degree of shortening). Only the second of these groups undergoing the relevant intervention is described here. The scoring system used to assess treatment outcome uses 6 factors relating to gait, ROM (hip and knee), dislocation (hip and knee), and residual shortening. Scores range from 6 to 14 points higher scores worse. 6 points = excellent 7-9 points = good ≥10 points = bad. No details provided of validation of this scale. Very little description given of the surgical technique.

Abbreviations used: PFFD, proximal femoral focal deficiency; ROM, range of movement			
Study details	**Key efficacy findings**	**Key safety findings**	**Comments**
Tönnis D (1997)[2] Study type: **Case series** Country: Germany n = 3 Study period: 1980 to 1992 Study population: PFFD, with some flexibility or displacement between the unossified part of the femoral neck and femoral head. Age: 4 days–5 weeks. Sex: not reported. Inclusion criteria: Not reported Technique: plaster cast in a squatting position to (110–120° of flexion, and 50° of abduction) for 3 months to aid fusion, followed by valgus osteotomy to bring the growth plate vertical to the load for maximal growth stimulation. Correction of retroversion to normal anterversion of the femoral neck. Follow-up: **2.3 to 8 years** Conflict of interest: Not reported.	**Patient 1** Patient with PFFD and some flexibility between the femoral head and metaphysic. The defect was almost corrected at 2.3-year follow-up, with an obvious varus deformity not apparent on the initial arthrogram, but no femoral anteversion. To increase growth a valgus osteotomy (with rotation of the femoral neck to 25° of anteversion) was performed at 3.2 years. At 8 years, radiographs showed excellent appearance of the proximal femur with mild coax vara. Limb length inequality was 8 cm. Femoral lengthening with a Wagner distractor was undertaken. **Patient 2** Initial radiographic assessment at 4 weeks of age showed proximal displacement of the femur in abduction and some mobility in the zone of lysis. The hip was immobilised in a cast for 10 weeks, a second arthrogram demonstrated fusion. The femoral neck was ossified to the region of the growth plate. Subsequent radiographs showed spontaneous development of the proximal femur. At 3.6 months of age the femoral length had increased significantly, but coax vara persisted. At 2.1 years excessive retroversion of the femoral neck was seen in relation to the position of the femoral condyles at the knee. During valgus osteotomy anteversion was increased to a normal angle. The limb length deficit compared to controls at 2.7 and 3.5 years was 6 cm and 6.5 cm, respectively. This constant level of deficiency will allow for lengthening to be performed later. **Patient 3** Arthrography at 9 days of age showed marked displacement between the femoral head and neck, and a hemispherical proximal femur. There was more mobility in this patient's hip than the other two patients. A hip spica cast in the squatting position was applied for 3 months. At 9 months the epiphyseal nucleus was almost normal in size, and the growth plate was developing regularly. There was marked coax vara, but the femur was becoming straighter. Valgus osteotomy with rotation of the femoral neck to normal anteversion was performed at 1.1 years, combined with a lateral acetabuloplasty on the opposite side for dysplasia and dislocation. By 2.2 years of age the epiphysis had a normal appearance and the femur had straightened. The limb length difference at 2.3 years was 7 cm, and amount that can be corrected without difficulty.		Method of case selection or case accrual not described. Surgical technique and staging of the intervention varies considerably between patients. Safety outcomes were not reported on.

Validity and generalisability of the studies

- There is a very limited evidence base in relation to numbers of patients reported in the peer-reviewed literature.
- There is considerable variation in the operative procedure from case to case.
- The degree of PFFD severity is not well described in the studies.
- There do not appear to be universally agreed or validated outcome measures to describe subjective efficacy parameters.
- No comparative data are available.

Specialist Advisers' opinions

Specialist advice was sought from consultants who have been nominated or ratified by their Specialist Society or Royal College. The advice received is their individual opinion and does not represent the view of the society.

Mr C Bradish (British Society for Children's Orthopaedic Surgery/British Limb Reconstruction Society), Prof. N Clarke (British Society for Children's Orthopaedic Surgery), Mr J Fernandez (British Society for Children's Orthopaedic Surgery/British Limb Reconstruction Society), Mr F Monsell (British Limb Reconstruction Society).

- Two of the Specialist Advisers considered this procedure to be a minor variation on an existing procedure and one considered it to be the first in a new class of procedures.
- Adverse events relating to the procedure known anecdotally or from the literature include significant hip and knee stiffness as a result of excessive lengthening, hip dislocation and recurrent deformity. The overall the result may be less good than would have been achieved prosthetically.
- Additional theoretical adverse events were thought to include avascular necrosis, failure of bone to unite, infection, nerve or vascular injury, poor limb function, recurrence of contractures and wound dehiscence.
- Key efficacy outcomes by which to evaluate this procedure were thought to be overall limb function, skeletal maturity and a reduction in the requirement for repeat operations.

- Surgeons should be competent in performing all the components of the procedure, and training is best undertaken by observing a surgeon versed in this technique.
- One adviser stated that the potential risks are of the same order as with any hip reconstruction in childhood.
- The main controversy in the management of the more severe forms of PFFD is whether to reconstruct all cases, or use selective reconstruction, and employ prosthetic management in the most severe cases.
- Outcomes and complications of the procedure should be reviewed.
- If the procedure were considered safe and efficacious, three Specialist Advisers thought that it would be offered only in less than 10 specialist centres, while one considered it impossible to predict the potential take-up at present.

Issues for consideration by IPAC

- The years of publication of the two studies are 1997 and 2002; however it would appear that the procedure is yet to be established as a safe and efficacious intervention (and that was the view taken at scoping stage).
- The search identified a number of other studies which focused on interventions for re-orientating limbs or distracting limbs to stimulate growth, but these were considered to be outside the scope of this overview because they did not address the combined procedure.

References

1 Fernandez-Palazzi F and Molina H. (2002) Proximal femoral focal deficiency. Results of conservative and surgical treatment, critical analysis and indications. Journal of Orthopaedics and Traumatology 3:97-101.

2 Tönnis D and Stanitski DF. (1997) Early conservative and operative treatment to gain early normal growth in proximal femoral focal deficiency. Journal of Pediatric Orthopaedics, Part B 6:59-67.

Appendix A: Additional papers on combined bony and soft tissue reconstruction for hip joint stabilisation in proximal focal femoral deficiency (PFFD)

The following table outlines the studies that are considered potentially relevant to the overview but were not included in the main data extraction table (table 2). It is by no means an exhaustive list of potentially relevant studies.

Article	Number of patients/follow-up	Direction of conclusions	Reasons for non-inclusion in table 2
Fixsen JA, Lloyd-Roberts GC (1974) The natural history and early treatment of proximal femoral focal dysplasia. The Journal of Bone and Joint Surgery 56: 86-95.	Case Report n=15 FU= not reported	Very few clinical data are reported	The procedures described do not directly fit within the intervention as defined in the scope. In many patients there was no soft tissue correction, and few procedures addressed the acetabular component of this condition. Few useful clinical outcomes are reported and the baseline characteristics of the patients are not well described. The paper largely focuses on prognosis based on radiological parameters.

Appendix B: Related NICE guidance for combined bony and soft tissue reconstruction for hip joint stabilisation in proximal focal femoral deficiency (PFFD)

Guidance	Recommendations
Interventional procedures	**Intramedullary distraction for lower limb lengthening. NICE interventional procedures guidance 197 (2006).** 1.1 Current evidence on the safety and efficacy of intramedullary distraction for lower limb lengthening does not appear adequate for this procedure to be used without special arrangements for consent and for audit or research. Although there is evidence of efficacy in lengthening the femur, evidence on its safety is inadequate. There is inadequate evidence on both efficacy and safety in lengthening the tibia. 1.2 Clinicians wishing to undertake intramedullary distraction for lower limb lengthening should take the following actions. • Inform the clinical governance leads in their Trusts. • Ensure that patients understand the uncertainty about the procedure's safety and efficacy in its use for lengthening the tibia and its safety in use for lengthening the femur, and provide them with clear written information. In addition, use of the Institute's information for patients ('Understanding NICE guidance') is recommended • Audit and review clinical outcomes of all patients having intramedullary distraction techniques for lower limb lengthening. 1.3 A number of devices are available for the procedure which may have different safety and efficacy profiles. The technology is continuing to evolve and clinicians should consider the choice of device on the basis of the most current available evidence. 1.4 Publication of safety and efficacy outcomes will be useful. The Institute may review the procedure upon publication of further evidence.
Technology appraisals	No relevant guidance
Clinical guidelines	No relevant guidance

Public health guidance	No relevant guidance

Appendix C: Literature search for combined bony and soft tissue reconstruction for hip joint stabilisation in proximal focal femoral deficiency (PFFD)

Database	Date searched	Version/files	No. retrieved
Cochrane Database of Systematic Reviews – CDSR (Cochrane Library)	02/10/2008	Issue 3, 2008	1
Database of Abstracts of Reviews of Effects – DARE (CRD website)	02/10/2008	N/A	9
HTA database (CRD website)	02/10/2008	N/A	0
Cochrane Central Database of Controlled Trials – CENTRAL (Cochrane Library)	02/10/2008	Issue 3, 2008	17
MEDLINE (Ovid)	02/10/2008	1950 to September Week 4 2008	371
MEDLINE In-Process (Ovid)	02/10/2008	October 01, 2008	23
EMBASE (Ovid)	02/10/2008	1980 to 2008 Week 39	466
CINAHL (NLH Search 2.0)	02/10/2008	1981 to Present	362
BLIC (Dialog DataStar)	29/09/2008	N/A	None found.
National Research Register (NRR) Archive	29/09/2008	N/A	None Found
UK Clinical Research Network (UKCRN) Portfolio Database	29/09/2008	N/A	None found
Current Controlled Trials *meta*Register of Controlled Trials – *m*RCT	29/09/2008	N/A	Anterior Cruciate Ligament (ACL) Reconstruction using two different types of Femoral Fixation i.e. Mitec Rigidfix femoral polylactide (PLA) cross pin and the Anthrex Bio Transfix implant
Clinicaltrials.gov	29/09/2008	N/A	None found

The following search strategy was used to identify papers in MEDLINE. A similar strategy was used to identify papers in other databases.

Searches

exp Bone Lengthening/

(ilizaro* adj3 technique*).tw.

(osteogenes$ adj3 distract$).tw.

callotas*.tw.

((femur* or femora* or thigh* or hip or hips) adj3 (reorientat* or re-orientat* or lengthen* or distract* or reconstruct* or re-construct* or realign* or re-align* or reposition* or re-position*)).tw.

((bony or bone*) adj5 (soft adj3 tissue*) adj5 (procedure* or surg* or correct*)).tw.

superhip*.tw.

(super adj3 hip*).tw.

or/1-8

Limb Deformities, Congenital/

Lower Extremity Deformities, Congenital/

Femur/ab

Thigh/ab

((femur* or femora* or focal* or thigh*) adj5 (short* or hypoplas* or abnormal* or anomal* or deficien* or discrepan* or deform* or dislocat* or dysplas*)).tw.

PFFD.tw.

Hip Dislocation, Congenital/

Hip Dislocation/

Hip Joint/ab [Abnormalities]

Hip/ab [Abnormalities]

((hip or hips) adj3 (hypoplas* or abnormal* or anomal* or deficien* or discrepan* or deform* or dislocat* or dysplas*)).tw.

or/10-20

9 and 21

animals/

humans/

23 not (23 and 24)

22 not 25

limit 26 to english language

from 27 keep 1-371