This paper reviews the current knowledge relating to the management of adult patients with congenital hip disease. Orthopaedic surgeons who treat these patients with a total hip replacement should be familiar with the arguments concerning its terminology, be able to recognise the different anatomical abnormalities and to undertake thorough pre-operative planning in order to replace the hip using an appropriate surgical technique and the correct implants and be able to anticipate the clinical outcome and the complications.

The use of ultrasound as a screening test in the newborn has allowed earlier diagnosis and treatment with a better prognosis for congenital disease of the hip. Orthopaedic surgeons who specialise in adult reconstructive surgery often face the problem of osteoarthritis secondary to congenital hip disease in adult patients. These cases are the result of late diagnosis and treatment in areas where early screening and treatment were not effective or in patients in whom treatment had failed in childhood.

Treatment of the young adult with congenital hip disease by periacetabular or intertrochanteric osteotomy is effective, but technically challenging. Acetabular osteotomy may be used in older patients with advanced degenerative disease secondary to congenital disease. The management of these patients with a total hip replacement (THR) presents difficulties because most are young with considerable demand on their implants and they may require complex reconstruction on both sides of the joint.

**Terminology and classification**

There is a need for an agreed terminology which covers the entire spectrum of congenital deformity of the hip and a classification of its types, both in infancy and adulthood, in order to improve communication, the planning of treatment and the evaluation of the results of different treatments. The term ‘developmental dysplasia of the hip’, which is widely used by orthopaedic surgeons throughout the world, does not reflect the congenital origin of the condition, while the indiscriminate use of the word ‘dysplasia’ does not accord with the variety of the underlying pathology. We favour the use of the term ‘congenital hip disease’ and its classification in infants as dysplasia, subluxation or dislocation. In adults, we have described three types of the disease, namely, dysplasia, low dislocation and high dislocation based on radiological and intra-operative criteria.

For a classification system to be useful in clinical practice, it should precisely describe the underlying pathological anatomy. It should also be able to predict the outcome of different methods of treatment and be simple and accurate to use. The classification proposed by Crowe, Mani and Ranawat is the most commonly used system for congenital hip disease. The main element of this system is the degree of displacement and the migration of the femoral head and describes four types of dislocation. The displacement is calculated on an antero-posterior pelvic radiograph by measuring the vertical distance between the inter-teardrop line and the junction of the femoral head and medial side of the femoral neck. The amount of dislocation is the ratio between this distance and the vertical diameter of the undeformed femoral neck. However, displacement and migration do not describe the underlying pathology, and this, in our opinion, limits the use of Crowe’s classification for the purpose of documentation only. Other commonly used classification systems are shown in Table I.

When dealing with these cases in the 1970s, before modern imaging techniques were available for pre-operative planning, we realised that not all of such hips were the same from an anatomical point of view and that different reconstruction techniques and...
materials were required for different cases. Notes on the intra-operative appearance of the hip were kept and compared with the radiographs in order to improve our pre-operative planning using conventional imaging. The whole spectrum of the disease was classified into three types (Fig. 1). In dysplasia (Fig. 2), the femoral head is contained within the true acetabulum, there is a superior segmental defect and the fossa is covered by an osteophyte which makes the acetabulum shallow. In a low dislocation (Fig. 3), the femoral head articulates with a false acetabulum which partially (to a varying degree) covers the true acetabulum. Apart from the superior segmental defect of the true acetabulum, there is an anterior segmental defect and a narrow opening of inadequate depth. Increased anteversion is seen in most cases. Occasionally there is a lack of posterior bone stock. In a high dislocation (Fig. 4), the femoral head migrates superiorly and posteriorly in relation to the hypoplastic, triangular true acetabulum. There is a segmental defect of the entire rim, and the acetabulum is shallow with a narrow opening. There is also an abnormal build-up of bone posterosuperiorly and excessive anteversion of the acetabulum. The iliac wing is hypoplastic and anteverted. The proximal part of the femur is normal in dysplasia, but the femoral neck is short in a low dislocation, and shorter still in a high dislocation with excessive anteversion (Fig. 5). The diaphysis is hypoplastic with excessive narrowing of the canal and thin cortices. Occasionally, there is a residual angular deformity of the proximal femur because of a previous osteotomy which can make the reconstruction much more challenging.

Table I. Details of classification systems for congenital hip disease

<table>
<thead>
<tr>
<th>Author/s</th>
<th>Types of increasing severity</th>
<th>Characteristic feature</th>
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<tbody>
<tr>
<td>Hartofilakis et al&lt;sup&gt;9-12&lt;/sup&gt;</td>
<td>Dysplasia Low dislocation B1 Low dislocation B2 High dislocation C1 High dislocation C2</td>
<td>Description of anatomical abnormalities</td>
</tr>
<tr>
<td>Crowe et al&lt;sup&gt;13&lt;/sup&gt;</td>
<td>I, &lt; 50%; II 50% to 75% III, 75% to 100% IV, &gt; 100%</td>
<td>Proximal migration/height of femoral head</td>
</tr>
<tr>
<td>Eftekar&lt;sup&gt;14&lt;/sup&gt;</td>
<td>Dysplasia Intermediate dislocation Intermediate</td>
<td>Degree of subluxation</td>
</tr>
<tr>
<td>Kerboul&lt;sup&gt;15&lt;/sup&gt;</td>
<td>Anterior Intermediate Posterior</td>
<td>Direction of subluxation</td>
</tr>
</tbody>
</table>

Diagram of the three main types of congenital hip disease in adults showing a) dysplasia, b) low dislocation and c) high dislocation.

Three-dimensional CT scan of a dysplastic left hip after removal of the femoral head. The segmental defect in the superior wall and the osteophyte covering the fossa are shown by arrows.
We have validated this classification system as have others\textsuperscript{16-18} and were satisfied that we could predict any local abnormalities from pre-operative radiographs. Later, we recognised, as a result of intra-operative problems with reconstruction, that both the low and the high dislocations could each be subdivided into two groups\textsuperscript{19}. In the B1 subtype of low dislocation there is extended cover of the true acetabulum by the false acetabulum and in the B2 subtype there is limited cover (Fig. 5). In the C1 subtype of high dislocation there is a false acetabulum high on the iliac wing and in the C2 subtype the femoral head lies within the abductor musculature (Fig. 6).

Pre-operative planning and reconstruction
Prior to surgery, an AP radiograph of the pelvis is obtained and in cases of low and high dislocation a CT of the involved hip is performed as well\textsuperscript{19}. The size and location of the segmental defects are estimated, as are the diameter and depth of the acetabulum (useful for the estimation of the required size of the acetabular component), the distribution of bone stock and the anteversion of the acetabulum and the femoral neck. The proximal femur is assessed using conventional radiographs and templates to estimate the type and size of the femoral component to be used for reconstruction.
The major technical difficulties encountered during a THR for congenital hip disease are reconstruction of the acetabulum in cases of low and high dislocation, implantation of the femoral component in a very narrow diaphysis and in patients with a residual angular deformity of the femur. We suggest that an osteotomy of the greater trochanter is included in the approach to the joint. This not only makes access easier, but may also restore the biomechanics if the trochanter is advanced distally thereby increasing the power of the abductor mechanism. Most surgeons currently use trochanteric osteotomy only in selected cases, mainly difficult primary and revision THRs. However, in our opinion, in patients with congenital dislocation of the hip, especially in low and high dislocations and in some stiff hips with dysplasia, it helps to avoid serious complications and gives a better result. For mechanical reasons, we favour placing the acetabular component at the level of the true acetabulum (Fig. 7). However, it is not always possible to achieve cover of the acetabular component with host bone at this level. In cases in which the reamed acetabulum can provide osseous cover of at least 80% (as it is estimated inter-operatively) we prefer an uncemented metal-backed acetabular component (Fig. 7). The size of these components is often small, 40 mm to 42 mm, and in order to avoid problems with thin polyethylene liners, some authors currently advocate the use of monobloc components or alternative bearing surfaces such as ceramic-on-ceramic (Fig. 8a). Good medium-term results have recently been reported using resurfacing arthroplasty in patients with dysplasia.
When it is not possible to use an uncemented acetabular component, the cotyloplasty technique is a good alternative.\textsuperscript{9,10} This involves medialisation of the acetabular floor by the creation of a cemented comminuted fracture of the entire medial wall, impaction of autogenous bone graft and the implantation of a small, cemented all-polyethylene acetabular component (Figs 8b and 8c).\textsuperscript{9,10}

Augmentation of superior segmental defects with structural autograft or allograft and the placement of the acetabular component in the anatomical position have been suggested (Fig. 7b).\textsuperscript{27,28} Although the short-term results of this technique are excellent, a high failure rate has been reported after approximately 12 years.\textsuperscript{28} This may be related to the complex pathological anatomy of the true acetabulum and the abnormal distribution of stresses combined with the unfavourable long-term behaviour of structural grafts. It has been suggested that this technique gives satisfactory results when at least 70\% of the acetabular component is supported by the host bone.\textsuperscript{29} High placement of the component in the region of the false acetabulum has also been proposed.\textsuperscript{27,30} The problem with this technique is that with the acetabular component at this level, the lever arm for the body-weight is much longer than that of the abductors, and causes excessive loading of the hip. Also, the shearing forces acting on the acetabular component can lead to early loosening. In unilateral cases a high acetabular component does not correct leg-length and leaves the patient with a limp.

We initially used a Charnley cemented femoral component exclusively for the reconstruction of the proximal
femur. Later, various small cemented components were also used (Figs 7 and 8). Several authors prefer uncemented femoral components (Figs 7 and 8). In our opinion, the principles and goals of uncemented fixation with optimal fit and fill of the canal, initial stability and adequate bone growth are not easily achieved in a narrow femoral canal with a thin cortex. One author (TK) currently performs the same procedure using a short cementless conical distal bearing component (Fig. 8a). Custom-made femoral components have also been used.

For hips with a high dislocation we favour reduction of the components by shortening the femur with progressive resection of bone from the femoral neck. We argue against leaving the greater trochanter in place and shortening the femur by subtrochanteric osteotomy because in most cases of high dislocation the greater trochanter lies above the centre of rotation of the femoral head. This makes trochanteric osteotomy and advancement essential. Subtrochanteric osteotomy may also lead to undesirable complications. Despite this, some surgeons continue to perform shortening osteotomies (either subtrochanteric or diaphyseal) with or without a trochanteric osteotomy. These techniques have been practiced over the years without clear indications for one or the other based on comparative results. It seems that detachment of the trochanter with shortening of the proximal metaphysis is indicated for proximal malformations which are difficult to treat; a mid-diaphyseal osteotomy is indicated when there is a concomitant severe valgus deformity of the knee, while a subtrochanteric shortening osteotomy has broader indications. When planning a shortening osteotomy, a CT of the pelvis and lower limbs is essential to measure the femora accurately and thus the true leg-length discrepancy. It has been found that in up to 30% of cases of low and high dislocation the affected femur is longer than that on the opposite side. Rarely, a two-level osteotomy is required to reduce the joint and equalise leg-lengths.

Clinical outcome
THR is the last resort for the treatment of an adult with osteoarthritis secondary to congenital hip disease. The young age of the patients combined with the altered morphology of the hip, and consequent lack of bone stock may result in a higher rate of a failure and a worse functional outcome. It should be stressed that the interpretation of the published results is difficult because most series include patients with many different types of hip disease.

Charnley and Feagin stated that THRs should be avoided in patients with congenital dislocation of the hip and inadequate bone stock. Despite this early discouragement, surgeons have attempted this procedure, developed their technique and reported their results. Overall, mid- and long-term failure rates vary from 7.7% to 44.8% in non-homogeneous series. In five homogeneous series with high dislocations there was a failure rate of 25% in a group of 87 patients at a mean follow-up of 10 years, of 17% in a smaller group of 28 patients at a mean follow-up of 9.4 years, of 10% in a group of 83 patients with a mean follow-up of eight years, and of 95% in a group of 52 patients with a mean follow-up of 12.3 years. In a recent study of 28 patients with Crowe type-IV hips treated with an uncemented implant and a subtrochanteric osteotomy, there was only one failure of the acetabular component but a high complication rate after a mean follow-up of 4.8 years. In our original series of 229 THRs, dysplastic hips performed as well as primary osteoarthritic hips with a survival rate of 90% at 15 years whereas in hips with low and high dislocation the survival rate was only 75% at the 15-year follow-up. Femoral components performed better in patients with a low dislocation while acetabular
components performed better in those with a high dislocation. Chougle, Hemmady and Hodgkinson reported similar findings for the long-term behaviour of the acetabular component in patients with a high dislocation.

Recent studies from centres in which these procedures are performed in sufficient numbers show satisfactory long-term clinical results with limited complication rates, even for patients with a high dislocation. Orthopaedic surgeons who treat adult patients with congenital hip disease should be familiar with its terminology, be able to recognize the different anatomical abnormalities, and be able to carry out thorough pre-operative planning, reconstruct the hip using the appropriate surgical technique and implants and finally be able to anticipate the clinical outcome and avoid complications.

Supplementary material
A figure showing anatomical abnormalities seen in high dislocation is available with the electronic version of this article on our website at www.jbjs.org.uk

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

References
CONGENITAL HIP DISEASE IN ADULTS


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