Total hip arthroplasty in patients with Down's syndrome

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Hip disease occurs in between 8% and 28% of patients with Down's syndrome, many of whom develop disabling pain. We have carried out total hip replacement in six adult patients (9 hips) with severe arthritis of the hip. The mean follow-up was 7.75 years (2 to 14). At the latest review, all had relief of pain and full hip function. Increasing longevity and a high incidence of hip disease in these patients suggest a greater role for total hip arthroplasty in the future.

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Down's syndrome is the most common chromosomal disorder in man. The incidence is 1 in 800 live births, but this varies with maternal age. There is an increased incidence of dislocation, dysplasia, slipped epiphysis, Perthes' disease, and avascular necrosis.¹

As the life expectancy for patients with Down's syndrome has increased, so has the presence of debilitating hip disease. This suggests a greater role for arthroplasty. Total hip replacement in such patients has been reported to give satisfactory results. We present the results of nine total hip arthroplasties in six patients with Down's syndrome.

Patients and Methods

We performed total hip arthroplasty in six patients (9 hips) with Down's syndrome and disabling arthritis. The mean age at surgery was 36 years (22 to 47). All patients were

mobile and lived at home or in a group residency at the time of operation.

The initial pathology leading to the need for surgery was avascular necrosis in one hip, slipped epiphysis with avascular necrosis in one, acetabular dysplasia with avascular necrosis in three and acetabular dysplasia in four (Table I).

Reamed bipolar prostheses were used in three hips because of an anticipated inability to follow a 'total hip' protocol. The remaining six hips were given standard total joint prostheses; two hips were braced and three were placed in spica casts during the postoperative period as a precaution. The posterior approach was used for each but the anatomical deformity required removal and replacement of the greater trochanter in three. The mean size of the acetabular component was 50.5 mm (44 to 55) and the mean diameter of the femoral shaft 10.5 mm (9 to 12). Eight of the nine arthroplasties were uncemented.

Results

There were no complications related to anaesthesia or wound healing. One patient (case 6) dislocated a reamed bipolar prosthesis on the fifth postoperative day. This required open reduction and no subsequent dislocation occurred. One patient (case 2) developed acetabular lysis which required revision 7 years and 4 months after the initial arthroplasty. This was attributed to malposition of the acetabular component, allowing impingement of the femoral neck against it. Polyethylene wear occurred leading to periacetabular lysis. Revision required removal and replacement of the greater trochanter which was secured with wires; these subsequently broke, allowing proximal migration of the trochanter. The patient has a painless limp (Fig. 1).

The mean length of follow-up was 7.75 years (2 to 14). Hip rating scores were considered to be unreliable because of the patients' limited comprehension. At the latest follow-up all patients were fully mobile and had no limitation of function due to their hips. None seemed to have any discomfort and all had good movement. Two had a mild limp associated with leg-length discrepancy and one a painless limp because of nonunion of the trochanter. All patients had equal leg lengths or less discrepancy than had been present before operation.

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Table I. Total hip replacement in patients with Down's syndrome

Case	Side	Date of surgery	Diagnosis	Age at surgery (yr)	Implant	Postoperative complications	Length of follow-up (yr)	Present status
1	R	9/80	SCFE* Avascular necrosis	22	Cemented mini-cad	None	14	Painfree Fully mobile
2	R	9/87	Avascular necrosis	28	Non-cemented AML-DePuy	Late acetabular revision	10.3	Painfree Fully mobile Limp
3	R	10/87	Acetabular dysplasia	28	Non-cemented osteonics	None	10.2	Painfree Fully mobile
	L	10/89	Acetabular dysplasia	30	Non-cemented osteonics	None	9.2	Painfree Fully mobile
4	L	11/88	Avascular necrosis Acetabular dysplasia	43	Non-cemented AML-DePuy	None	9.1	Painfree Fully mobile
	R	9/89	Acetabular dysplasia	44	Non-cemented AML-DePuy	None	8.3	Painfree Fully mobile
5	R	5/93	Acetabular dysplasia	44	Non-cemented Bipolar-DePuy	None	4.6	Painfree Fully mobile Shoe lift
	L	1/96	Avascular necrosis Acetabular dysplasia	47	Non-cemented Bipolar-DePuy	None	2	Painfree Fully mobile
6	L	6/94	Avascular necrosis Acetabular dysplasia	42	Non-cemented Bipolar--DePuy	Dislocation day 5 after operation	3.5	Painfree Fully mobile Shoe lift

* slipped capital femoral epiphysis

Fig. 1a

Fig. 1b

Fig. 1c

Fig. 1d

Fig. 1e

Fig. 1f

Case 2. Radiographs showing a) the normal right hip 15 months before surgery, b) avascular necrosis of the right hip ten months before operation, c) the right hip immediately before operation, d) the valgus position of the acetabular component, e) marked acetabular lysis after 68 months and f) wire breakage which had resulted in proximal migration of the trochanter after acetabular revision.

Fig. 2a

Fig. 2b

Fig. 2c

Fig. 2d

Fig. 2e

Fig. 2f

Case 5. Radiographs showing a) preoperative acetabular dysplasia and severe osteoarthritis of the right hip, b) the right hip with a reamed bipolar prosthesis, c) the left hip at the time of arthroplasty of the right hip, d) subluxation of the left hip two months after arthroplasty of the right side, e) the left hip at 31 months after right hip arthroplasty with progressive dysplasia, avascular necrosis, and osteoarthritis and f) the bilateral reamed bipolar prostheses, 53 (right) and 21 (left) months after operation.

Discussion

Hip disease occurs in between 8% and 22% of patients with Down's syndrome. There is an increased incidence of dislocation, dysplasia, slipped epiphysis, Perthes' disease and avascular necrosis. Shaw and Beals¹ reviewed 228 hips of patients between 10 and 43 years of age and found radiological abnormalities in 8%. Hresko, McCarthy and Goldberg² assessed 130 adult hips and found radiological abnormalities in 22%. Cristofaro, Donovan and Cristofaro³ reported an incidence of osteoarthritis of 10% in an adult population with Down's syndrome. Increasing longevity in patients with Down's syndrome suggests that debilitating hip disease will become more common.⁴⁻⁷

A study of the anatomy of the hip in this condition showed an increase in acetabular depth, a decrease in acetabular anteversion and a roof which is more horizontal than normal. This should create a mechanically stable hip, but the presence of a lax capsule and an increased range of movement, especially in external rotation, appears to make

the hip vulnerable to the development of arthritis.² Each of these relatively young patients had radiological evidence of severe degenerative changes, but the pathophysiology which led to disabling arthritis was not always clear. Spontaneous dislocation of the hip occurs in children with Down's syndrome, and can result in acetabular dysplasia.⁶ None of the patients in our series had a history of hip dislocation; several had rapid progression of hip disease in mid-adult life. When this occurred, surface osteonecrosis and focal arthritis of the femoral head were seen in hips with acetabular dysplasia. We suspect that ligamentous and capsular laxity allows the head to sublux with consequent dysplasia and arthritis. The cause of the high incidence of avascular necrosis (5 of the 9 hips in our series) is not clear (Fig. 2).

Preoperative evaluation of patients with Down's syndrome should take into account several medical conditions which are associated with the disorder. These include patellar hypermobility, instability of the upper cervical spine, cardiac anomalies, susceptibility to infection,⁷⁻¹³ and the early development of Alzheimer's dementia.

Atlantoaxial subluxation occurs in 10% to 30% of patients with Down's syndrome. Preoperative lateral flexion/extension radiographs of the cervical spine will reveal this, and any other abnormality.⁷⁻⁹ Congenital cardiac anomalies are present in about 40% of such patients.¹² They also have abnormalities of the immune system and are known to suffer an increased incidence of infection. No surgical infection occurred in our series nor has infection been reported in other patients with Down's syndrome who have had a hip arthroplasty. Careful postoperative monitoring is necessary to detect infection of the urinary tract and venous thrombosis; these patients may not report symptoms. Uncertain comprehension and temperament limit their ability to comply with postoperative instructions. Bracing, casting, and reamed bipolar prostheses were used in our series with good results.

Reamed bipolar arthroplasty has been advocated as a primary treatment of arthritis and also for patients with recurrent dislocation.¹³⁻¹⁵ The technique involves reaming the acetabulum to the subchondral bone and inserting a bipolar cup of the same size as the largest reamer. This prevents movement of the acetabular component and creates a stable hip. We have successfully used this technique in patients with recurrent dislocation, severe mental incapacity, dystonia musculorum, and myotonic dystrophy as well as in patients with Down's syndrome.

The complications which required surgical revision in our series could have been avoided. The early dislocation of a bipolar prosthesis (case 6) might have been prevented if the acetabulum had been reamed more deeply. The late periacetabular lysis (case 2) may not have occurred if the position of the acetabular component had been less open.

There have been only a few reports in the literature of patients with Down's syndrome who have had hip replacement. Skoff and Keggi¹⁶ described eight total replacements in five patients. The mean age at operation in their series was 46 years. All patients had acetabular dysplasia and subluxation of the femoral head. Excellent results were achieved with no evidence of infection, loosening, or dislocation at a mean follow-up of 4.3 years. Most of those reported in the literature and eight of nine patients in our series have been treated with ingrowth femoral components.

Replacement arthroplasty is a satisfactory treatment for

severe hip disease in patients with Down's syndrome. Preoperative preparation should take into consideration the anatomy of the socket, any associated medical conditions and the degree of postoperative compliance possible. Modification of the surgical technique and postoperative protocol may be necessary to achieve satisfactory results. The high incidence of hip disease in patients with Down's syndrome suggests a greater role for total hip replacement in the future.

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