CASE REPORT

Bilateral total hip replacement in arthrogryposis multiplex congenita

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SUMMARY

The authors present a case of bilateral total hip replacements (THRs) in a 56-year-old patient with arthrogryposis multiplex congenita (AMC). The considerations for the perioperative period and the outcome are discussed. Preoperative planning included an anaesthetic review and availability of fiberoptic intubation due to poor mouth opening. Perioperatively, contractures can make positioning and exposure difficult but in this case a standard posterior approach was taken. Particular attention was given to soft tissue balancing given the theoretical risk of dislocation. There were no perioperative complications. Postoperatively there has been improvement in pain and hip scores but the patient has failed to return to work. Objective improvements in range of motion (ROM) have not been made. This experience suggests THR is a safe and effective treatment for osteoarthritic hip pain in patients with AMC but patients should be informed that ROM is unlikely to improve.

BACKGROUND

Arthrogryposis multiplex congenita (AMC) is a rare congenital disease. It affects 1 in 30 000 live births.¹ The exact pathophysiology is unknown. It is associated with upper limb deformities, foot and ankle deformities, scoliosis, knee contractures and hip conditions.

The hips are affected in approximately 30–80% of cases.^{2–5} The most common hip conditions are contractures, dislocations and subluxations.² The long-term mobility of these patients varies greatly and almost half the patients in the studies published were ambulating independently at the time of the study.^{2–5} Others needed assistance in the form of crutches or walking aids.²

The published results of performing total hip replacement (THR) in this group of patients are mixed.^{6–8} There are technical difficulties involved in performing the operation. The soft tissue issues could theoretically increase the risk of dislocation and other complications although the literature is limited to a small number of case reports.

We present the results of our experience to add to the limited published evidence available to guide surgeons caring for patients with AMC. It is important that patients with AMC understand not only the benefits but also the limitations and risks associated with a hip replacement in this cohort.

CASE PRESENTATION

We present a case of a 56-year-old man, a bartender, with a diagnosis of AMC, who underwent bilateral THRs. His medical history included hypertension and mild depression.

Congenital dislocation of the hips was noted at birth. The diagnosis of AMC was made at the age of 15 years. As a child, the patient had undergone a number of soft tissue procedures on his hips and ankles. He underwent bilateral-staged Chiari osteotomies at the age of 27 and 29 years. The knee joints were relatively spared.

Non-hip-related orthopaedic issues included bilateral ankle fusions and bilateral carpal tunnel decompressions in 2007 and 2010. Currently, he is being evaluated for development of subtalar arthritis bilaterally. He wears bilateral ankle and foot orthosis splints.

INVESTIGATIONS

Preoperative ranges of motion and hip scores are shown in [table 1](#). Preoperative X-rays showed bilateral osteoarthritis (OA) secondary to developmental dysplasia, with bilateral Crowe Grade (CG) 3 changes.

TREATMENT

The patient had a primary left THR in July 2008. A standard posterior approach to the hip was used—piriformis and short external rotators were detached, and capsulotomy was performed. Evidence of developmental dysplasia (CG3) was noted as well as grade 4 osteoarthritic changes in the femoral head and acetabulum. The implant used included a modular femoral system and uncemented cup—S-ROM 18 B small proximal sleeve and standard stem, standard neck, and a Pinnacle 36 mmID×58 mmOD cup with metal on metal bearing surfaces. The capsule was repaired with interrupted sutures. Piriformis and short external rotators were attached with two transosseous sutures. Stability was checked through normal range of motion (ROM).

The patient underwent a right THR in December 2008. A standard posterior approach was performed identical to the previous left THR. The iliopsoas was additionally detached from its femoral insertion to allow adequate intraoperative exposure of the acetabulum. Again, signs of significant dysplasia (CG3) and OA were noted. Impaction grafting of the superior lateral acetabulum was performed to increase coverage. Closure was performed similarly to that of the left THR. Both procedures were carried out under spinal anaesthetic with fiberoptic intubation available on standby.

OUTCOME AND FOLLOW-UP

The postoperative course was uneventful. At 7-year follow-up, the patient is pain free. He has no history of dislocation. [Table 1](#) compares his

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Table 1 Range of motion and hip score preoperatively and at latest follow-up

	Right preoperative	Right postoperative	Left preoperative	Left postoperative
Abduction	11–15	35	<10	30
Adduction	11–15	11–15	<10	11–15
Internal rotation	0	15	0	20
External rotation	<10	<10	<10	11–15
Flexion	40–45	40	66–75	45
Extension	0	<10	10	11–15
Harris hip score	34.2	72.4	31.95	72.4

preoperative ROM and hip scores, with range and scores measured in June 2015. Postoperative X-rays are shown in [figure 1](#). There are no differences in hip ROM in the immediate postoperative period to those at the latest follow-up.

DISCUSSION

This case represents a successful case of hip replacement performed in a patient with AMC. The patient no longer experiences any pain. He had no complications from the procedure. ROM was not greatly changed from his preoperative state.

There are a number of studies and publications on the management of hip dislocations in children with AMC. Dislocations can be unilateral or bilateral. Historically, emphasis was placed on treating unilateral dislocation aggressively due to risk of developing scoliosis and pelvic obliquity while bilateral dislocations could be treated conservatively.⁴ Recently, there has been a trend towards surgical management of all dislocations in these patients.^{9 10}

There is little published in the literature regarding management of these patients as adults. THR in this patient posed a number of challenges. Owing to the patient's underlying condition, there were specific considerations made to preoperative planning, the intraoperative course and expectations of long-term outcome.

Figure 1 Recent pelvic radiograph showing bilateral hip replacements in situ with no evidence of complication.

Owing to poor mouth opening, related to the AMC fiberoptic, intubation was necessary as a standby measure in case of prolonged surgery or an ineffective spinal requiring a switch to general anaesthesia.¹¹ Although it was not needed in this case, it is important that the facility is available on site when planning such a case. It is also an important consideration to be aware of when treating these patients in a trauma setting.

Patients with AMC have a tendency for presenting with fibrosis and contractures. Adequate patient positioning in these patients may be an issue. The knees of the patient in question were spared from the disease and, in this case, the positioning was straightforward and he was easily placed in the standard lateral position.

Exposure using a posterior approach was difficult in both cases and the right THR required a psoas tenotomy both for exposure of the acetabulum and to improve postoperative hip extension. Although we could be confident that the pain would be relieved, soft tissue balancing in these patients is a great concern. Disrupting soft tissues with unpredictable healing theoretically would increase the risk of postoperative dislocation.

While the patient has not returned to work, he is otherwise fully independent and mobilises with one stick outside of the home. ROM has not shown substantial objective improvement from preoperative values (improvement in internal rotation and abduction, and no improvement in flexion) and attempting hip replacement for this goal would not be recommended. This finding is in line with the published literature. Cameron *et al*⁷ equally found no improvement in ROM. Leonard *et al*⁶ described positive results in a patient who underwent a THR, and who had AMC and an ipsilateral above knee amputation. Fisher *et al*⁸ reported on a case of bilateral knee replacements, THR and contralateral revision THR in a patient, resulting in functional and pain improvements but no improvement in objective hip ROM. Patients with AMC who are being considered for THR should be made aware of this likely outcome in preoperative counselling as well as in the consenting process. In spite of these drawbacks, our patient reports subjective increase in ROM and this is reflected in improved hip function scores.

As previously mentioned, although there is literature on the management of these patients with a normal life expectancy, only one previous study has described THR in this cohort. The theoretical reason for this may be the low rate of OA pain in these patients due to decreased ROM and also decreased mobility. Surgeons may be reluctant to consider THR in these patients, considering the previously described complicating factors. Our case shows that, in those patients experiencing OA pain in the setting of AMC, THR may be offered as a safe and effective operation for relieving pain. THR should not be recommended with the intention of increasing ROM.

Learning points

- ▶ Total hip replacement is a safe and effective operation for relieving pain in patients with arthrogryposis multiplex congenita (AMC) and hip osteoarthritis.
- ▶ THR does not improve range of motion (ROM) in patients with AMC.
- ▶ Functional hip scores show improvement in post-THR in patients with AMC.
- ▶ A standard posterior approach can be used, but additional soft tissue release may be required to gain adequate exposure to perform the surgery.

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Competing interests None declared.

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REFERENCES

- 1 Thompson GH, Bilenker RM. Comprehensive management of arthrogryposis multiplex congenita. *Clin Orthop Relat Res* 1985;194:6–14.
- 2 Yau PW, Chow W, Li YH, *et al.* Twenty-year follow-up of hip problems in arthrogryposis multiplex congenita. *J Pediatr Orthop* 2002;22:359–63.
- 3 Carlson WO, Speck GJ, Vicari V, *et al.* Arthrogryposis multiplex congenita. A long-term follow-up study. *Clin Orthop Relat Res* 1985;194:115–23.
- 4 Huurman WW, Jacobsen ST. The hip in arthrogryposis multiplex congenita. *Clin Orthop Relat Res* 1985;194:81–6.
- 5 St Clair HS, Zimble S. A plan of management and treatment results in the arthrogryposis hip. *Clin Orthop Relat Res* 1985;194:74–80.
- 6 Leonard M, Nicholson P. Total hip arthroplasty in a patient with arthrogryposis and an ipsilateral above knee amputation. *Hip Int* 2010;20:559–61.
- 7 Cameron HU. Total joint replacement in multiplex congenita contractures: a case report. *Can J Surg* 1998;41:245–7.
- 8 Fisher KA, Fisher DA. Total hip and knee replacement in a patient with arthrogryposis multiplex congenita. *Am J Orthop (Belle Mead NJ)* 2014;43:E79–82.
- 9 Akazawa H, Oda K, Mitani S, *et al.* Surgical management of hip dislocation in children with arthrogryposis multiplex congenita. *J Bone Joint Surg Br* 1998;80:636–40.
- 10 Asif S, Umer M, Beg R, *et al.* Operative treatment of bilateral hip dislocation in children with arthrogryposis multiplex congenita. *J Orthop Surg (Hong Kong)*. 2004;12:4–9.
- 11 Steinberg B, Nelson VS, Feinberg SE, *et al.* Incidence of maxillofacial involvement in arthrogryposis multiplex congenita. *J Oral Maxillofac Surg* 1996;54:956–9.

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