Hip Dysplasia in Mucopolysaccharidosis Type IVA (Morquio A Syndrome) Treated by Proximal Femoral Valgization Osteotomy: A Case Report

Josephine Berger-Groch[1,2], Martin Rupprecht[2], Ralf Stuecker[2], Nicole Muschol[3], Sandra R Breyer[1,2]

Learning Point of the Article:

Arthrography is essential in the treatment of hip dysplasia. A valgus osteotomy in cases with hinge abduction can be a successful solution.

Abstract

Introduction: Morquio A syndrome or mucopolysaccharidosis type IVA (MPS IVA) is a progressive lysosomal storage disorder caused by an N-acetylgalactosamine-6-sulfatase deficiency. The abnormal metabolism of glycosaminoglycans among other medical problems leads to various skeletal disorders caused by a dysfunction of endochondral ossification of epiphyseal cartilage. Severe hip dysplasia is common and can lead to pain and impaired mobility.

Case Report: We report on a 15-year-old girl suffering from MPS IVA. At the age of 5 years, hip pain and a reduced walking distance were described for the 1sttime. At the age of 9 years, acetabulofemoral dysplasia associated with genuavalga was diagnosed. After pre-operative assessment of the hips including plain radiographs, magnetic resonance imaging, and arthrography with dynamic testing a valgization osteotomy of the proximal femur in combination with a shelf acetabuloplasty was performed. The patient was followed for 6 years with a stable hip joint and without any sign of redislocation.

Conclusion: Some treatment strategies of hip dysplasia in patients with MPS IVA are described in the literature. The techniques used for congenital hip dysplasia, varisation of the femur in combination with Pemberton, Salter, or shelf acetabuloplasty, are widely reported. Nevertheless, resubluxations were described in some cases. The well-known surgical procedure with valgization of the proximal femur is not reported in literature for MPS IVA patients. In our opinion, dynamic testing with arthrography should strongly be considered for this particular problem before surgical intervention. Pathology-related decisions should be made under consideration of the different surgical techniques.

Keywords: Arthrography, hip dysplasia, Morquio, mucopolysaccharidosis, shelf acetabuloplasty, valgization.

Introduction

Morquio A syndrome or mucopolysaccharidosis type IVA (MPS IVA) is a progressive lysosomal storage disorder caused by an N-acetylgalactosamine-6-sulfatase deficiency. The abnormal metabolism of glycosaminoglycans among other medical problems leads to various skeletal disorders caused by a dysfunction of endochondral ossification of epiphyseal cartilage. Severe hip dysplasia is common and can lead to pain and impaired mobility[1,2]. The techniques used for congenital hip dysplasia, varisation of the femur in combination with Pemberton, Salter, or shelf acetabuloplasty, are widely reported for MPS patients. Nevertheless, resubluxationswere described after this technique [3].The well-knownsurgical approach with valgization[4]and shelfacetabuloplasty in cases with hinge abduction is not described for MPS IVA patients in literature before.

Case Report

We report on a 15-year-old girl suffering from MPS IVA. Diagnosis was suspected due to an older affected sibling and

Access this article online

Website: www.jocr.co.in

DOI: 2250-0685.1208

Author's Photo Gallery


Dr. Josephine Berger-Groch


Dr. Martin Rupprecht


Dr. Ralf Stuecker


Dr. Nicole Muschol


Dr. Sandra R Breyer

[1]Department of Trauma-, Hand-, and Reconstructive Surgery, University Medical Center, Hamburg-Eppendorf, Martinistraße, 52, 20246 Hamburg, Germany,
[2]Department of Pediatric Orthopedics, Children's Hospital Altona, Bleickenallee 38, 22763 Hamburg, Germany,
[3]Department of Pediatrics, University Medical Center, Hamburg-Eppendorf, Martinistraße, 52, 20246 Hamburg, Germany.

Address of Correspondence:
Dr. Josephine Berger-Groch,
Department of Trauma-, Hand-, and Reconstructive Surgery, University Medical Center, Hamburg-Eppendorf, Martinistraße, 52, 20246 Hamburg, Germany.
E-mail: j.berger@uke.de

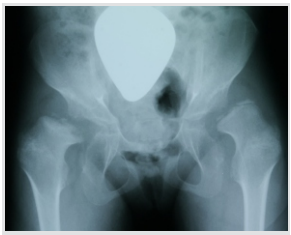

Figure 1: Pre-operative plain anteroposterior radiography of the pelvis. Patient's age: 9.3 years.

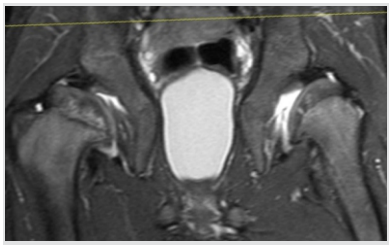

Figure 2: Pre-operative magnetic resonance imaging of the hip joints (T2).

confirmed at 9 months of age by enzymatic as well as genetic testing. Her cognitive and motor development was age appropriate. Independent walking was achieved at the age of 15 months, and the girl was clinically asymptomatic at the time of diagnosis. At the age of 5.0 years, the girl complained about pain in hips and knees. The first orthopedic assessment at the interdisciplinary outpatient clinic of our institution was done at the age of 9.3 years. At this time, the patient weighed 19 kg and was 114 cm tall. Mobility (e.g., walking distance and ability to climb stairs) was already restricted. There was a symmetrically reduced range of motion of the hip joints of EXT/FLEX 0°/0°/120°; IR/ER 30°/0°/40°; and ABD/ADD 40°/0°/30°. In addition, genuavalga was present without restriction of passive range of motion of the knees. A systematic orthopedic workup was carried out. Atlantoaxial instability and myelopathy were excluded by cervical magnetic resonance imaging (MRI). Standing radiographs of the lower extremities showed a severe valgus deformity. The tibiofemoral angle was 26° on the right and 24° on the left side; the mechanical lateral distal femoral angle was abnormal on both sides (right 74° and left 75°). Furthermore, a plain pelvic radiography was performed (Fig. 1). This revealed flattened and nearly unossified femoral epiphyses. The neck of both femora was short and severe lateralization was seen with interruption of the Menard-Shenton line. The migration index according to Reimers [5] was 67% on the left side and 59% on the right side. The center-edge angle (CEA) of Wiberg [6] was –33° on the left side and –28° on the right side. MRI confirmed the subluxation of the hip joints. The cartilaginous and labral roof were completely deformed and proximally displaced. Intraarticular effusion was present in both hip joints (Fig. 2). Temporary epiphyseodesis (8plates) of the medial distal femur and medial proximal tibia on both sides was carried out to correct the severe valgus deformity. For a better understanding of the hip dysplasia, an arthrography was performed during the same anesthesia. Dynamic testing of the hips revealed incongruency in abduction

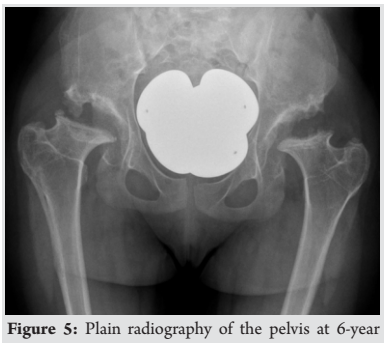

Figure 5: Plain radiography of the pelvis at 6-year follow-up. Patient's age: 15.6 years.

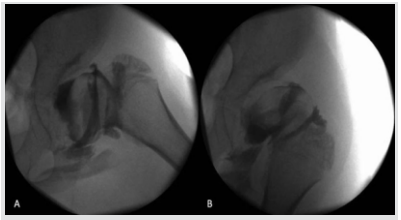

Figure 3: Intraoperative arthrography of the left hip; (a) in max. abduction, hinge abduction, (b) in 20° adduction.

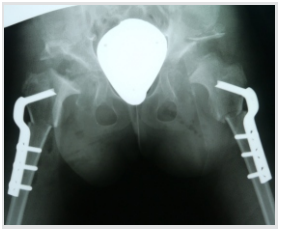

Figure 4: Post-operative plain radiography of the pelvis in spica cast.

on both sides. Hinge abduction with medial pooling and displacement of the labrum was obvious. In 20° of adduction, the femoral head and the acetabulum were congruent without any medial pooling (Fig. 3). In adduction, the labrum was horizontal without displacement. However, the bony roof of the acetabulum was too short in relation to the size of the femoral head. The center collum diaphyseal angle was reduced to 115° on both sides. To restore, joint congruency and coverage a valgization of the proximal femur of 20° in combination with a shelf acetabuloplasty were performed (Fig. 4). Post-operative immobilization was done with a spica cast in 20° abduction for 6 weeks followed by an in-house rehabilitation for 4 weeks. Due to a persistent flexion contracture of the right knee, which progressed during further growth, an anterior epiphyseodesis of the distal femur was performed at the age of 10.8 years. A year later (age: 11.9 years), the hardware was removed. At 12.0 years of age, enzyme replacement therapy (ERT) was initiated. At the last follow-up (age: 15.6 years), the patient weighed 35 kg and was 141 cm tall. Walking abilities were still limited - however, she was able to ambulate without crutches for small distances. The range of motion was still symmetrically reduced with EXT/FLEX 0°/0°/120°; IR/ER 40°/0°/30°; and ABD/ADD 30°/0°/20°. The supine anteroposterior pelvic radiograph revealed stable hips with a CEA of 19° on the left side and 33° on the right side. The migration index of Reimers improved to 25% on the left side and 23% on the right side (Fig. 5).

Discussion

Severe hip dysplasia is common in MPS IVA patients and can lead to pain and immobility[7].Clinical studies showed that hematopoietic stem cell transplantation is no effective therapeutic option for MPS IVA patients [8].IntravenousERTwas approved for the treatment of MPS IVA in 2014[9]. ERT has led to increased quality of life for MPS patients and has improved endurance as measured by an increased walking distance. However, acquired skeletal deformities do not improve after ERT and long-term outcome data are not available yet [2]. Due to an incomplete endochondral ossification of the epiphyseal cartilage, patients with MPS IVA typically present with a flattened and fragmented femoral head. In combination with a shallow and short

acetabulum, the deformity leads to a progressive dislocation of the hip and often rapid degeneration of the joint over time[10].The commonly described surgical procedure, as recommended for developmental dysplasia of the hips, is a proximal femoral varus osteotomy combined with acetabuloplasties, for example, a Pemberton, Salter, or Shelf procedure[1].Nevertheless,resubluxationswere described after this technique [3].There is no literature about the hinge abduction phenomenon in this patient and the resulting treatment options. The valgization of the proximal femur is a widely performed technique for congenital and acquired hip pathologies[4] but still not described for hinge abduction in patients with MPS IVA. Dhawale et al.[3] reported on a retrospective reviewof13 non-surgically treated MPS IVA patients and 15 surgically treated MPS IVA patients (average age 6.8±3.4 years)with hip dysplasia and found superior results after surgeryin these patients. All conservatively treated joints were subluxated or dislocated during follow-up. In the group treated with proximal femoral varus derotation osteotomy (VDRO) in combination with a shelf acetabuloplasty, no recurrent hip subluxation was found, whereas patients treated with Salter or Pemberton acetabuloplasties and/or VDRO alone needed revision surgery with shelf acetabuloplasty. The same complication was addressed by Al Kaissi et al.[11]in a casereportof a 6-year-old boy with MPS IVA. 5years after bilateral VDRO and Salter osteotomy a posterior dislocation of the left hip occurred. In contrast to these results, Kanazawa et al.[12]presented an MPS IVA patient with a good outcome after bilateral varus osteotomies at the age of 18years and a follow-up of 5 years. However, this patient was only mildly affected. The radiographs showed bilateral ossification defects in the main load-bearing area of the femoral head without signs of sub- or dis-location. Borowski et al.[13]recommended two-dimensional computed tomography (2D-CT) in MPS IVA patients before surgery. The 2D-CT scans of the hip demonstrated global acetabular deficiency with a predominantly anterosuperior dysplasia. There is no documented case of valgus osteotomy of the proximal femur in MPS IVA patients in the literature. Furthermore, pre-operative arthrographies are not described as a routine diagnostic tool. Plain radiography can lead to misunderstanding of the pathology. MRI delineates the cartilaginous coverage and should be performed before surgery. CT scans might give comparable information but should be avoided in young children due to radiation.

Conclusion

In summary, we want to focus on the importance of an arthrography although plain radiography is not able to show the cartilage part of the hip joint. Surgical intervention for hip dysplasia in MPS IVA patients should be individualized based on clinical examinations, pre-operative imaging including plain radiography, MRI, and dynamic arthrography. In our opinion, understanding of the dynamic function of the hip is essential for the surgical decision-making process. Our successful case with a 6-year follow-up illustrates that the well-known and widely performed valgus osteotomy in cases of hinge abduction in MPS IVA is analternative, which should be considered preoperatively.

Clinical Message

Arthrography is essential in the treatment of hip dysplasia. Based on the result, a valgus osteotomy in cases with hinge abduction can be a successful solution.

References

1. White KK, Jester A, Bache CE, Harmatz PR, Shediac R, Thacker MM, et al. Orthopedic management of the extremities in patients with morquio A syndrome. J Child Orthop 2014;8:295-304.
2. Hendriksz CJ, Berger KI, Giugliani R, Harmatz P, Kampmann C, Mackenzie WG, et al.International guidelines for the management and treatment of morquio A syndrome. Am J Med Genet A 2015;167A:11-25.
3. Dhawale AA, Thacker MM, Belthur MV, Rogers K, Bober MB, Mackenzie WG, et al. The lower extremity in morquio syndrome. J Pediatr Orthop 2012;32:534-40.
4. Sidler-Maier CC, Reidy K, Huber H, Hierauer S, Ramseier LE.LCP 140° pediatric hip plate for fixation of proximal femoral valgisation osteotomy. J Child Orthop 2014;8:29-35.
5. Reimers J. The stability of the hip in children. A radiological study of the results of muscle surgery in cerebral palsy. Acta Orthop Scand Suppl 1980;184:1-00.
6. Wiberg G. Studies on dysplastic acetabula and congenital subluxation of the hip joint. Acta Chir Scand Suppl 1939;83:7-135.
7. Cooper GA, Southorn T, Eastwood DM, Bache CE. Lower extremity deformity management in MPS IVA, morquio-brailsford syndrome: Preliminary report of hemiepiphysiodesis correction of genu valgum. J Pediatr Orthop 2016;36:376-81.

8. Tomatsu S, Sawamoto K, Alméciga-Díaz CJ, Shimada T, Bober MB, Chinen Y, et al. Impact of enzyme replacement therapy and hematopoietic stem cell transplantation in patients with morquio A syndrome. Drug Des Devel Ther 2015;9:1937-53.
9. Hendriksz CJ, Burton B, Fleming TR, Harmatz P, Hughes D, Jones SA, et al. Efficacy and safety of enzyme replacement therapy with BMN 110 (elosulfase alfa) for morquio A syndrome (mucopolysaccharidosis IVA): A phase 3 randomised placebo-controlled study. J Inherit Metab Dis 2014;37:979-90.
10. Tomatsu S, Mackenzie WG, Theroux MC, Mason RW, Thacker MM, Shaffer TH, et al. Current and emerging treatments and surgical interventions for morquio A syndrome: A review. Res Rep Endocr Disord 2012;2012:65-77.
11. Al Kaissi A, Klaushofer K, Grill F. Progressive acetabular dysplasia in a boy with mucopolysaccharoidosis type IV A (Morquio syndrome): A case report. Cases J 2008;1:410.
12. Kanazawa T, Yasunaga Y, Ikuta Y, Harada A, Kusaka O, Sukegawa K, et al. Femoral head dysplasia in morquio disease type A: Bilateral varus osteotomy of the femur. Acta Orthop Scand 2001;72:18-21.
13. Borowski A, Thacker MM, Mackenzie WG, Littleton AG, Grissom L. The use of computed tomography to assess acetabular morphology in morquio-brailsford syndrome. J Pediatr Orthop 2007;27:893-7.

Conflict of Interest: Nil
Source of Support: Nil

Consent: The authors confirm that Informed consent of the patient is taken for publication of this case report

How to Cite this Article

Berger-Groch J, Rupprecht M, Stuecker R, Muschol N, Breyer S R. Hip Dysplasia in Mucopolysaccharidosis Type IVA (Morquio A Syndrome) Treated by Proximal Femoral Valgization Osteotomy: A Case Report. Journal of Orthopaedic Case Reports 2018 Sep-Oct; 8(5): 50-53.