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Clinical Studies

Cranio-cervical decompression associated with non-instrumented occipito-C2 fusion in children with mucopolysaccharidoses: Report of twenty-one cases

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ABSTRACT

Background: Mucopolysaccharidosis (MPS) is a multisystemic storage disorder of glycosaminoglycan deposits. Infiltration of the dura mater and supporting ligaments caused spinal cord compression and consecutive myelopathy, especially at the cranio-cervical junction (CCJ). Craniocervical instability and posterior decompression often raise the problem of fixation in children. The main purpose of this paper was to report the result of an original technique of occipito-cervical arthrodesis using a cranial halo-cast system in pediatric population.

Methods: We recorded 21 patients with cervical myelopathy. All of them had spinal cord decompression by enlargement of the foramen magnum, C1 laminectomy, and occipito-C2 fusion using corticocancellous bone graft. Only one child has an extended laminectomy from C1 to C3. The occiput-C2 arthrodesis was stabilized by the cranial halo-cast system. This immobilization was performed preoperatively and kept for three months then switched to rigid cervical collar. Clinical assessment, including the Goel grade and mJOA, radiographs and magnetic resonance imaging were performed before surgery. The occipito-cervical arthrodesis was controlled by standard X-rays and CT scan.

Results: According to the type of mucopolysaccharidosis, the patients were divided into MPS type I: n= 3, II: n=7, IV: n=11. The mean age of patients at surgery was 6.76 years. All mucopolysaccharidoses cases required a foramen magnum decompression by craniectomy, C1 laminectomy and occipito-C2 arthrodesis. As major complications, a child had immediate post-operative paraplegia due to spinal cord ischemia. The postoperative follow-up ranged from 1.5 to 4 years, with an average of 3.3 years. The average preoperative mJOA score was 8.9, and it improved to 14 points at the last follow-up.

Conclusions: Satisfactory fusion and good clinical results were obtained with the 2-stage approach to CCJ anomalies.

Introduction

Mucopolysaccharidosis (MPS) is a multisystemic storage disorder of glycosaminoglycan (GAG) deposits [1]. Infiltration of the dura mater and supporting ligaments causes craniocervical instability and spinal cord compression. This leads to myelopathy, particularly at the craniocervical junction (CCJ) [2]. There are several challenges with regards to the surgical management which requires fusion in growing infants. The small size of the patients, the bone anatomy, the inherent ligamentous laxity and the growth potential of the vertebrae are responsible for the difficulties in treatment. Posterior spinal cord decompression and cranio-C2 arthrodesis often raise the problem of fixation in children with MPS. The technique of two-stage approach for unstable pediatric CCJ anomalies with a cranial halo-cast and delayed occipitocervical fusion has been barely studied in the pediatric population [3].

The main objective of this work was to report the result of cranio-cervical decompression and to describe an original technique of occipito-cervical arthrodesis using the cranial halo-cast system in the pediatric population with MPS.

FDA device/drug status: Not applicable.

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Table 1
Modified Japanese Orthopaedic Association Scoring System [4].

Score	Description
Motor function—upper extremity	
1	Paralysis
2	Fine motor function massively decreased
3	Fine motor function decelerated
4	Discrete weakness in hands or proximal arm
5	Normal function
Motor function—lower extremity	
1	Unable to walk
2	Need walking aid on flat floor
3	Need handrail on stairs
4	Able to walk without walking aid, but inadequate
5	Normal function
Sensory	
1	Apparent sensory loss
2	Minimal sensory loss
3	Normal function
Bladder function	
1	Urinary retention
2	Severe dysfunction
3	Mild dysfunction
4	Normal function
Maximum score: 17	

Material and methods

We carried out a retrospective study of 21 cases of children with MPS. This study concerned patients operated between January 2017 and December 2020 in a Pediatric and Adolescent Orthopedic Center. The management of this condition was multidisciplinary. It included a pediatric metabolic center, a pediatric radiology center, a pediatric anesthesia center, a pediatric neuro-surgery center, and a pediatric orthopedic center.

Demographic, clinical and radiological data of patients with MPS were collected during follow-up. The diagnosis of spinal cord compression at the level of the CCJ and the instability factors of the hinge were assessed by cerebrospinal MRI in all the children. We performed a PubMed literature research using the keywords: mucopolysaccharidosis, children, craniocervical stenosis, craniocervical decompression, non-instrumented occipitocervical fusion. The selected articles were in English published in indexed journals.

Assessments to evaluate pathology of the CCJ

The Clinical evaluations included the modified Japanese Orthopaedic Association score (mJOA) (Table 1) [4] and the Goel grading system (Table 2) [5] before and after setting up the Cranial halo-cast system, 3 months after surgery and at the final follow-up visit. For the MRI evaluation, both T1 and T2-weighted sequences were performed. The presence of a compression of the spinal cord was deduced from the sagittal and axial T2-weighted images. On MRI, spinal cord compression is defined as a lack of spinal fluid at the level of CCJ and/or basilar impression of the spinal cord. The T2-weighted images were also assessed to detect increased signal intensity, suggesting myelomalacia.

Table 2
Goel clinical grading system [5].

Grade	Description
1	Independent and normally functioning
2	Walks on own but needs support/help to carry out routine household activities
3	Walks with minimal support and requires help to carry out routine household activities
4	Walks with heavy support and unable to carry out routine household activities
5	Unable to walk and dependent for all activities

Surgical management

The operation was always performed by the same surgeons (Pediatric orthopedic surgeon Pr Nessib .M.N and Associate Pr Zairi .M and Pediatric neurosurgeon Associate Pr Bouali .S) using the same procedure. The surgery was performed in two-stages approach.

First stage: cranial Halo-cast placement

It consisted in fixing and stabilizing the CCJ with a halo-cast (Fig. 1) under general anesthesia. The distraction components were built from the distractors, hinges, and connectors of an Ilizarov apparatus. The relay between the cranial halo and the cast vest was made by hinges which allow flexion/extension of the cervical spine. The fixation of the cranial halo was carried out using 6 needles including 2 frontal, 2 parietal and 2 occipital. Second stage: Cranio-cervical decompression and Occiput-C2 posterior fusion:

7 to 10 days later, we performed the second stage. After fiberoptic intubation, the patient installation is done in prone position. The use of a halo-vest for positioning is of great importance to secure the cranium [6]. The head was held by the cranial halo. The cervical spine is flexed using the hinges to facilitate surgery (Fig. 2).

An incision was made from the protuberantia occipitalis externa right to the spinous process of C5. Soft tissue dissection concerns the muscle insertions at the level of the occipital protuberance and the posterior arches of C1 and C2.

The supraspinous and interspinous ligaments C2 - C3 were preserved so as not to inducespinal instability, or even junctional kyphosis. Then, C1 arch laminectomy, foramen magnum decompression, and thickened atlanto-occipital membrane dissection of the dura mater were performed (Fig. 3). No further stabilization of the CCJ was performed. After spinal cord decompression, the cervical spine is set to a neutral position. The patient was still in the prone position, we clinically identify the posterior iliac crest, an incision of the skin and soft tissues until we have the outer cortex of the iliac bone, then the bone graft was taken in block. Two corticocancellous bone rods, taken from the posterior iliac crest (Fig. 4A), werefixed between the occiput and the C2 arch (Fig. 4B). The bone, from C1 laminectomy and enlargement of the foramen magnum, is mixed with auto-graft for arthrodesis (Fig. 4C).

Postoperative care

Extubation was only performed when the patient was fully awake without neurological impairment, due to the technical difficulties of reintubation. The cranial halo-cast device was kept in place for 12 weeks after the second surgery. It was then removed with local anesthesia at an outpatient visit, and replaced by a rigid cervical collar for 3 months.

Statistical analysis

Effectivity analysis was carried out for clinical outcome (Goel grade and modified Japanese Orthopaedic Association score).

Student t test was used for analysis of quantitative variables. Findings were considered to be statistically significant when $P < 0.05$.

Complications

Postoperative complications resulting from the first procedure and the second surgery were reviewed.

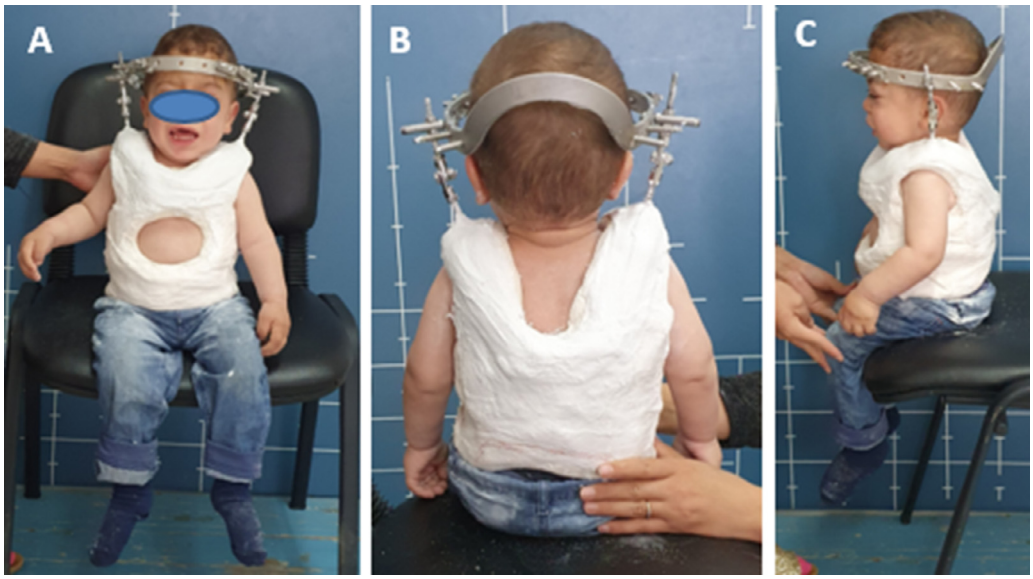

Fig. 1. Clinical photograph from the front (A), from behind (B) and from the side (C) showing the halo-cast in place preoperatively in a 3-year-old child with Morquio's disease.

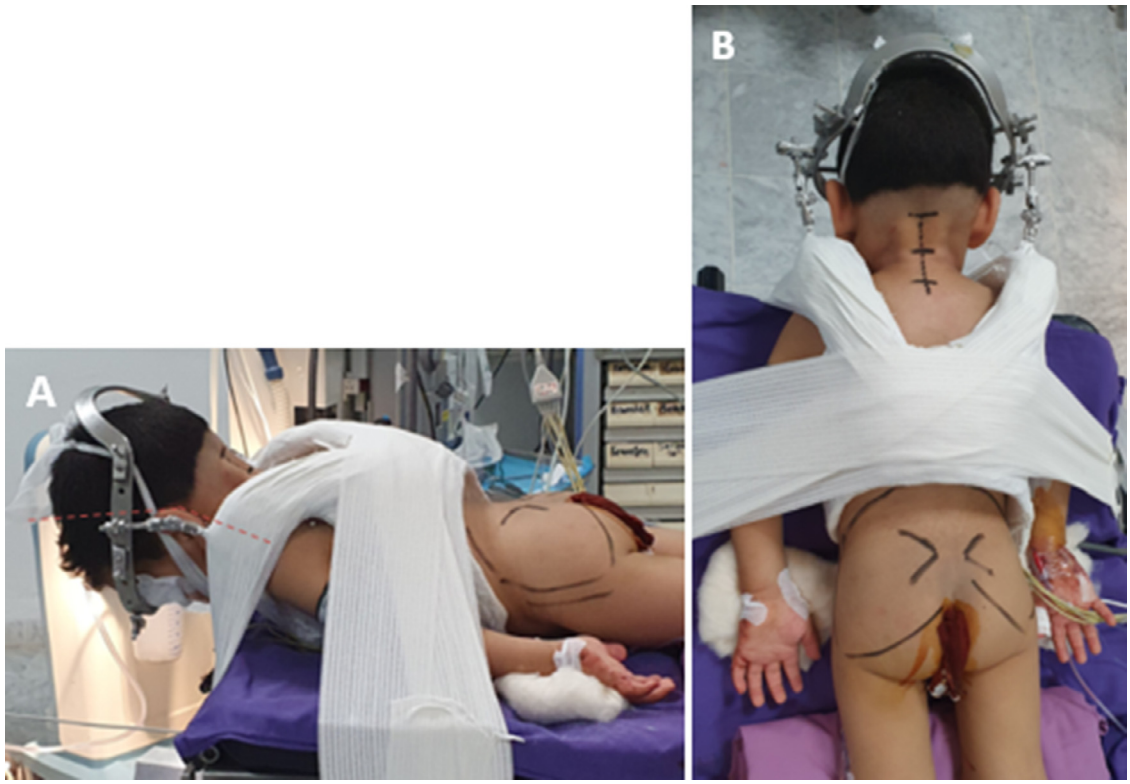

Fig. 2. Positioning the patient in the prone position and fixing to the surgical table (A: lateral view, B: view from above). The red dotted line on the lateral view indicates the degree of flexion of the cervical spine.

Results

The study included 21 patients, 12 males and 9 females. The median age at the time of decompression surgery was 6.76 years (ranges, from 3 to 15 years). According to the type of mucopolysaccharidosis, the patients were divided into MPS type I n= 3, II: n=7, IV: n=11. The postoperative follow-up ranged from 1.5 to 4 years, with an average of 3.3 years.

At the time of surgery, the MRI was objectified a compression of the spinal cord at the level of the CCJ with lack of the cerebrospinal fluid. A basilar impression was objectified in 7 patients. The main stenotic level was in foramen magnum and C1 for 20 childrens (Fig. 5) and extended for C3 for one patient.

Patients were stayed in the hospital for the whole procedure. The duration of hospitalization was 13 days on average (ranges, from 10 to 21 days) in the absence of post-operative complications. Between the

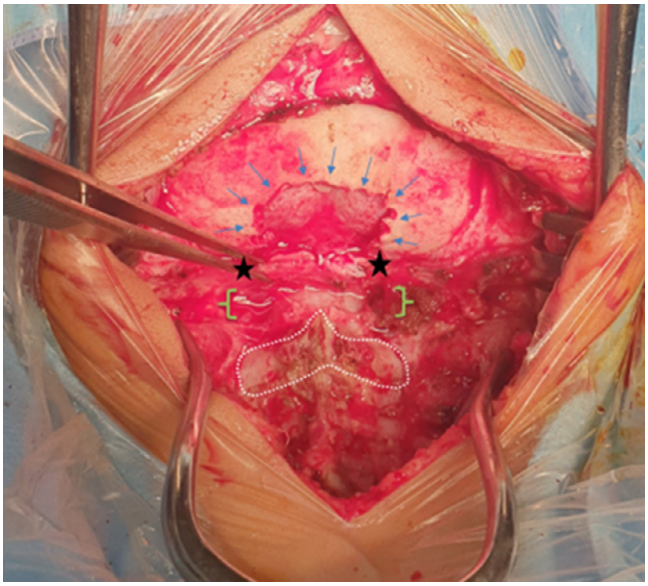

Fig. 3. Intraoperative photograph showing craniocervical decompression: the blue arrows delimit the level of the craniotomy, the 2 black stars delimit the level of the occipito-atloid ligament, the braces in green delimit the laminectomy of C1 and the pointed white line delimits the posterior arc of C2.

2 stages of surgery, the child was kept for progressive distraction at a rate of 4 mm/day for decapitation of the spine and for neurological monitoring. All patients underwent surgery according to the procedure aforementioned. The average operating time was 130 minutes (ranges, from 120 to 150 minutes). Mean intraoperative blood loss was 220 mL (range, 200–250 mL).

Clinical outcome analysis

After the first stage of surgery, we were noted an improvement in neurological status in 8 patients. Three of them, who had incomplete tetraplegia, were recovered partial motor skills in all four limbs. The mean mJOA score before surgical treatment was 8.9 points (ranges, 6–13 points) in the 21 patients. It improved to 14 points (ranges, 4–17 points) at the last follow-up. There were not statistically significant differences in the improvements between the different etiologies according to the postoperative mJOA grade ($P > 0.05$). 80% of the patients (17/21) were improved neurologically in the year following the operation with an improvement in motor and sensory status objectified by mJOA score and Goel grade. At final follow-up, their neurologic condition was preserved.

The mean Goel grade before both procedures was 3.19 (ranges, 2–5). It decreased to 1.52 at the last follow-up. Like mJOA score analysis, there were not statistically significant differences in the improvements between the different etiologies according to the postoperative Goel grade ($P > 0.05$).

The occipito-C2 arthrodesis was controlled by standard radiographs. The first radiological assessment carried out three months postoperatively makes it possible to objectify the shadow of the cortico-cancellous graft. The second radiological assessment, carried out six months postoperatively, was objectified occipito-C2 arthrodesis in 76 % of cases (16/21) and no signs of occipito-cervical instability in all patients (Fig. 6). At one year postoperative, CT scan was objectified arthrodesis in all operated children (Fig. 7) allowing a better analysis of the quality of arthrodesis in the three dimensions.

At the mean follow-up (3.3 years), the outcome of the 2-stage procedure was considered optimal in all 21 cases with 100 % successful occipito-C2 arthrodesis and 80 % improvement in sensory-motor status.

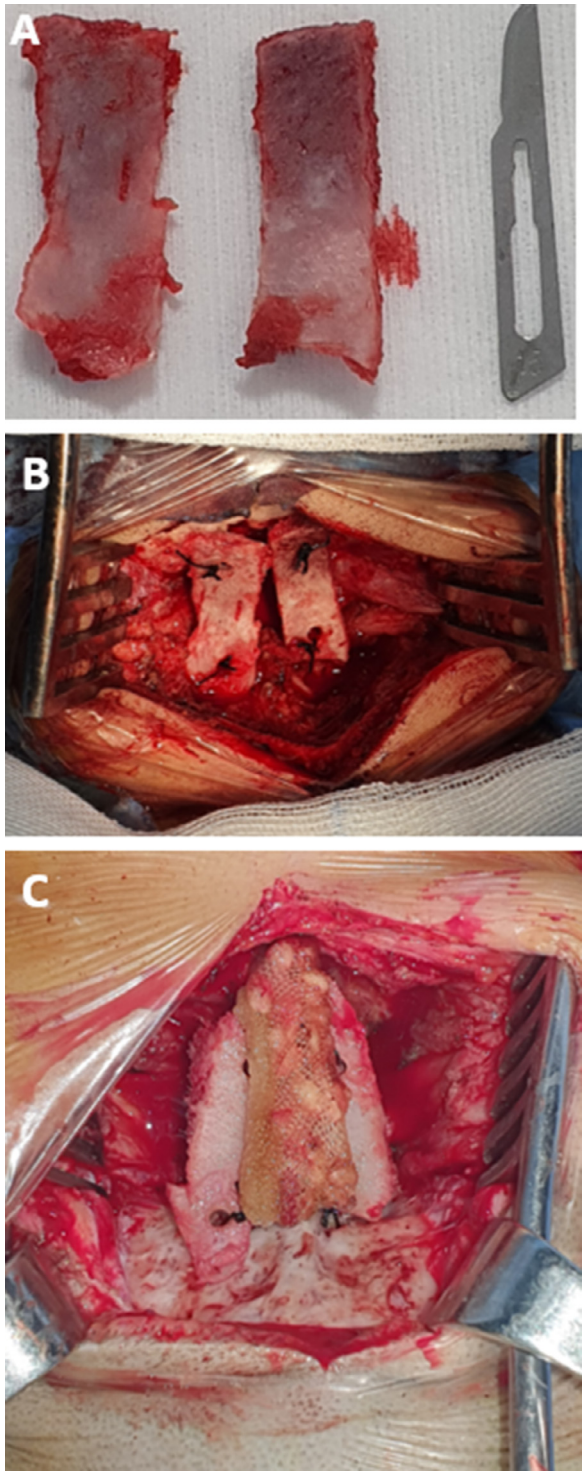

Fig. 4. Intraoperative photograph showing corticocancellous graft procedure : two corticocancellous bone rods, taken from the posterior iliac crest (Fig. 3A), are fixed between the occiput and the C2 arch (Fig. 3B). The product of C1 laminectomy and enlargement of the foramen magnum is associated for arthrodesis (Fig. 3C).

Complications

Postoperative complications resulting from the first procedure and the second surgery were reviewed.

Local complications: There was a case of loosening of the cranial halo following a fall, which was put back in place under general anesthesia. A superficial sepsis at the level of the needles was objectified in

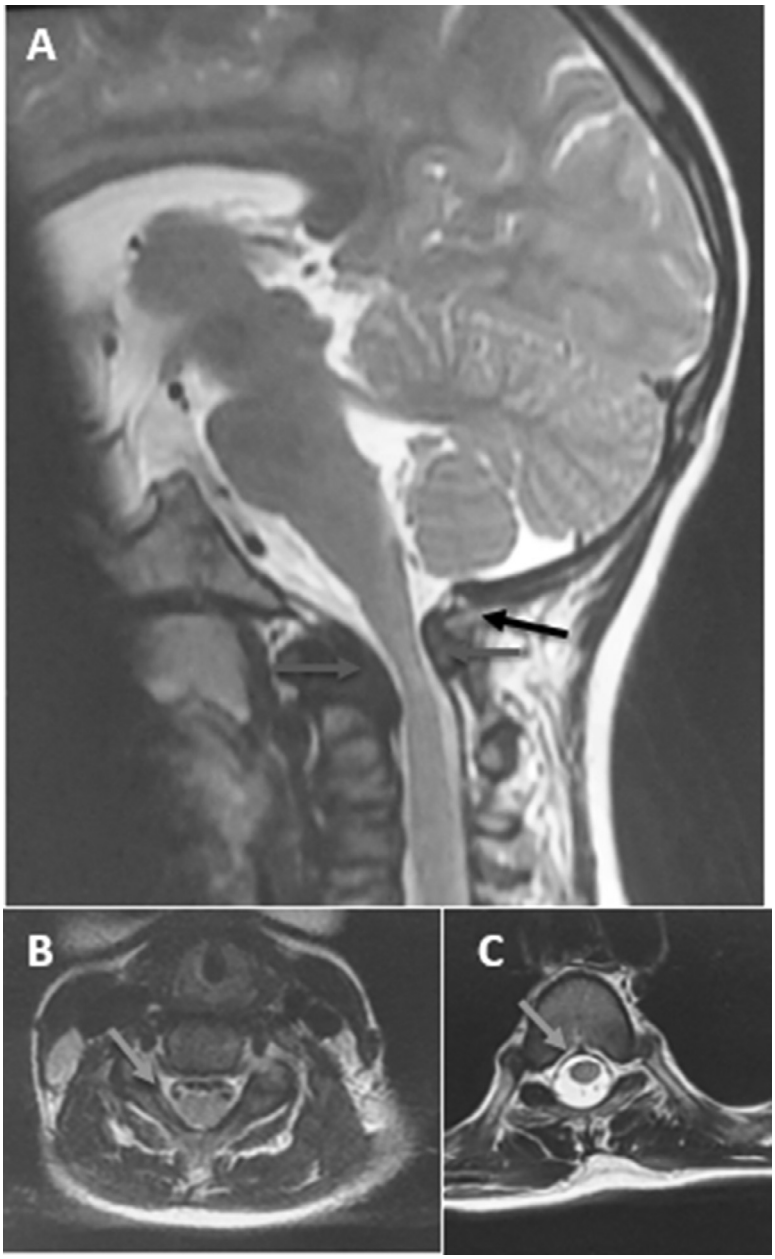

Fig. 5. MRI of a 5-year-old child showing in sagittal section (A) the basilar impression secondary to the protrusion of the scale of the occipital bone in the foramen magnum (arrow) and the compression of the spinal cord at the level of C1 (rarrow). The axial section passing through C1 (B) showed the absence of cerebrospinal fluid around the spinal cord (orange arrow) while the axial section passing through the thoracic vertebrae (C) showed a normal appearance of cerebrospinal fluid around the spinal cord (arrow).

3 children, which evolved well after local care and antibiotic therapy. No cerebrospinal fluid leaks were seen after removal of the needles. At the level of the cervical surgical site and the graft donor area, we didn't note any sepsis or disunity of the suture.

General complications: after cranio-C1 decompression and occipito-C2 fusion in Morquio's disease in a 5-year-old child who had flaccid tetraparesis preoperatively, a worsening of the neurological state was noted following the occurrence of thoracic spinal cord ischemia, leading to complete paraplegia. There were no cases of vertebral artery injury.

Discussion

Mucopolysaccharidosis disturbs the normal systemic endochondral and membranous bone growth after birth. This leads in various skeletal impairments, known as multiplex dysostosis, including the spine [7–9]. The involvement of the CCJ was almost constant. It is manifested by C1-C2 instability [10] and spinal cord compression [11] at the level of the foramen magnum and C1. The attention of the examine should be mainly focused on cervical pain or torticollis, suggestive of CCJ instability, progressive impairment of autonomous ambulation, postural instability, and fatigue and progressive weakening of upper limbs [11]. These elements are suggestive of a slow and progressive myelopathy [12]. Hence, the surgical management must have as an objective the decompression of the spinal cord and the fusion of the CCJ. A successful fusion depends on meticulous surgical planning, selection of appropriate instrumentation, optimizing graft for fusion, and postoperative immobilization [13]. The occurrence of these disorders in young children is a challenge for the surgeon [14]. MRI, including active dynamic flexion and extension scans, is the most powerful imaging technique to detect spinal cord compression at the CCJ level in MPS patients [11–15]. In Giussani's series, the diameter of the spinal canal was reduced at the level of the CCJ in 40% of patients. He was demonstrated that the most severe spinal canal stenosis and cord compression were seen in MPS IV (33%) and MPS VI (50%) [11]. It was recommended to perform neurological monitoring and spinomedullary MRI every year [16]. In addition to cervical stenosis, multilevel subaxial stenosis has been reported, especially in MPS VI, mainly due to posteriorly protruding in-

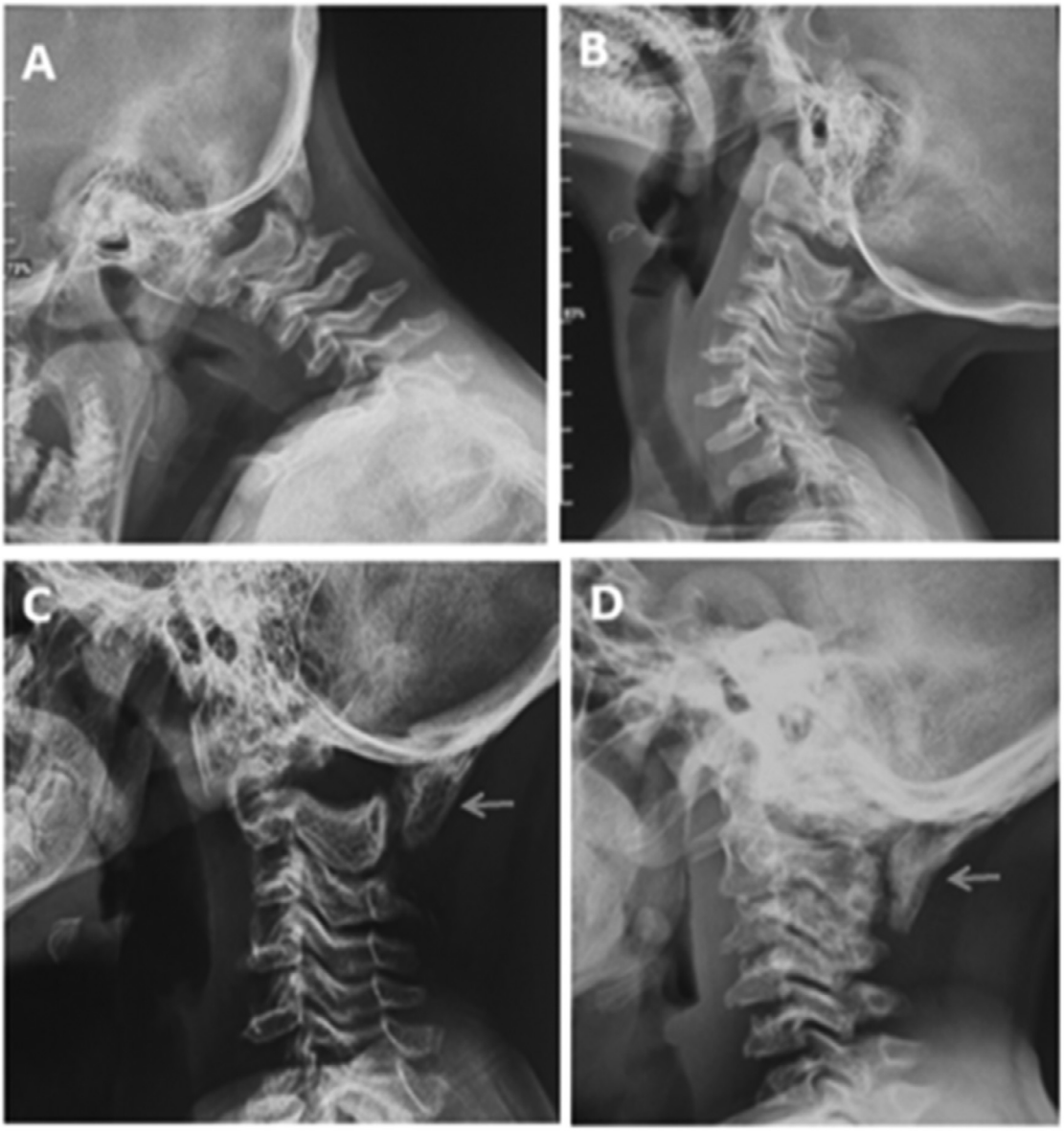

Fig. 6. Lateral radiograph of the cervical spine performed 6 months postoperatively on a 10-year-old child (A, B and C) and another 15-year-old (D). The orange arrows showed the appearance of the corticocancellous graft at 6 months. Dynamic flexion (A) and extension (B) X-rays demonstrated the stability of the occipito-cervical junction.

tervertebral discs, thickened dura, and hypertrophy of the ligamentum flavum [17].

The therapeutic management strategy was different according to the authors. Surgery should take into account the growth of an immature spine in young children [14]. The presence of neurological signs or myelopathies on the MRI was an absolute indication for surgery [18,19]. For other authors, the presence of neck pain or torticollis, signs of instability of the CCJ, was also an indication for surgery [3,20]. In our series, the indication for surgery was posed in the presence of a neurological deficit, which was associated with signs of instability and spinal cord compression on MRI. Intraoperative reduction employing a single surgical approach has been preferred by some authors [21–23], and several techniques have been applied for this purpose in pediatric patients [3,21,23]. Cranio-C2 stabilization with internal osteosynthesis material can interfere with placement of the bone graft for occipito-cervical fusion. Moreover, delicate and small bony structures may not withstand rigid instrumentation due to excessive mobility, poor bone purchase at the time of instrumentation, and still developing ossification centers with cartilaginous interpositions [24]. This technique has been improved over time [25,26]. However, these techniques were not devoid of risks, in particular the failure of the intraoperative reduction C0-C1-C2 with a fusion in a suboptimal anatomic situation. Hence, the uses of two-stage approach. The concept of preoperative traction techniques for reduction of these malformations was not new. This technique has also been improved over time. From bed traction [27] to halo-gravity traction [28], we were led to progressive traction by halo-cast or halo-vest.

Performing spinal cord decompression and craniocervical fusion using the halo-cast has several advantages. This was important for a variety of reasons. It allowed progressive occipito-vertebral decapitation, complete immobilization of the head and neck during positioning. Anatomically and functionally, occipitovertebral fusion by halo-cast allows the avoidance of growth disturbance, preservation of motion, and prevention of iatrogenic injury to the ossification zone and the smooth lamina, which may be caused by wires or screws [29].

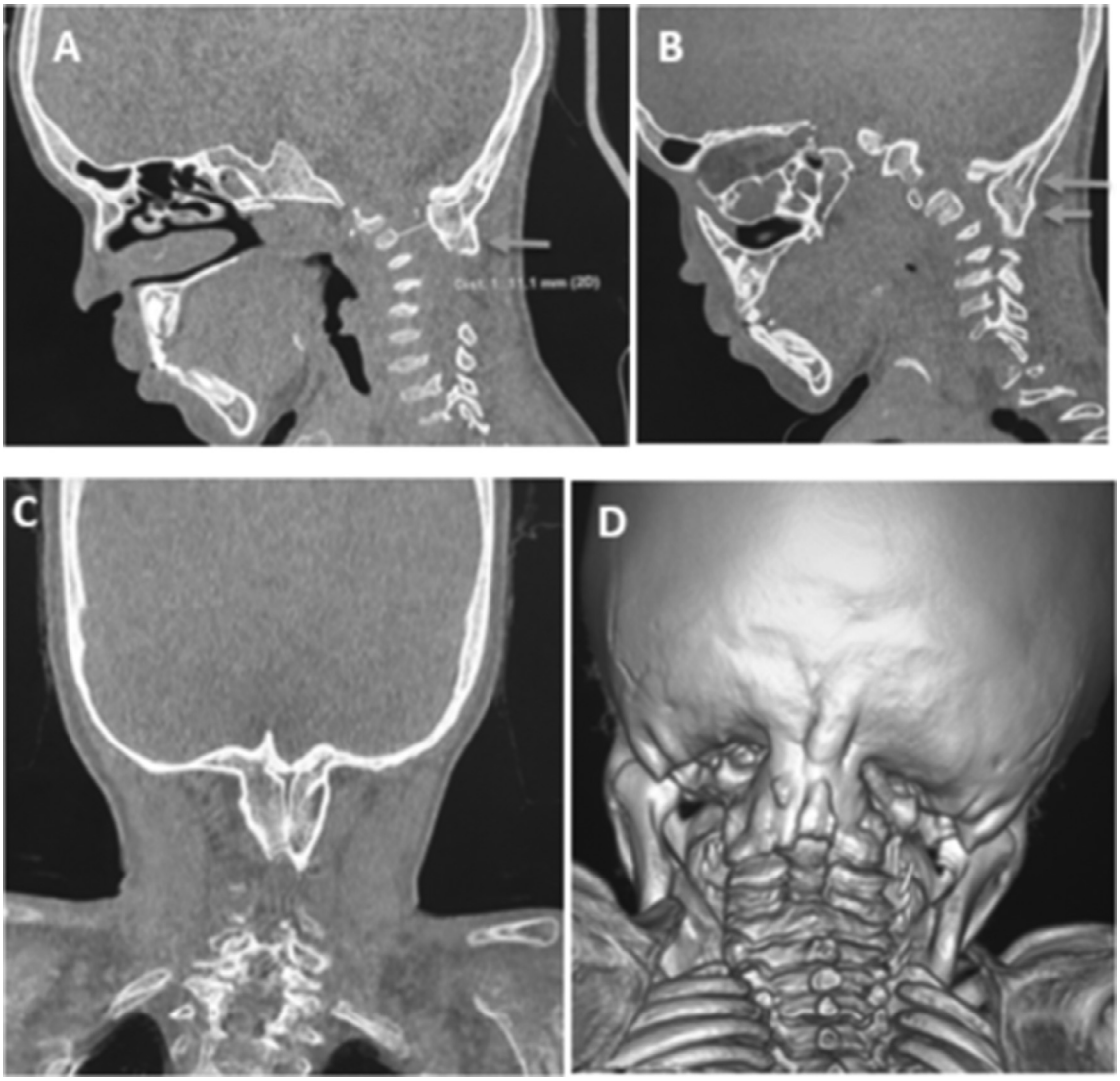

Fig. 7. CT scan of the cervical spine performed 12 months postoperatively in a 4-year-old child. The sagittal section (A) showed the normal dimension of the foramen magnum and the cervical canal. The orange arrows showed the aspect of the occipito-C2 arthrodesis in sagittal (A and B), coronal (C) sections and in posterior view in 3D (D).

Bone arthrodesis was optimally promoted by autologous grafts that have the advantages of being osteogenic, osteoinductive, and osteoconductive [30]. The choice of bone graft was crucial [14]. Corticocancellous graft was mechanically stronger than pure cancellous bone and was thereby ideally suited to support multiaxial movements across the occipitocervical articulation [18]. The autologous iliac crest graft was the most commonly used [31]. In our series, we were used two corticocancellous grafts in the form of a bagette and fixed with non-absorbable suture at the level of the occiput and the C2 posterior arch. As with many authors [32], the bone dust was mixed with auto-ograft to ensure better fusion. Authors have used rib autografts [14,33] and vascularized occipital bone graft [34], with good results. It was recommended utilization of absorbable sutures to avoid foreign material and minimize the risk of deep infections [35].The number of subaxial cervical levels to be included in an occipitocervical fusion was based on the child's diagnosis, clinical presentation, and radiographic findings. Children with isolated occipitocervical instability usually undergo an occiput–C2 fusion [36].

The halo vest was a simple and safe immobilization tool that was widely used for treatment of craniocervical lesions [37–40]. Preoperative traction stabilizes the CCJ in children with MPS and contribute to maintain the reduced position of the basilar invagination and the atlantoaxial dislocation [41].

The progressive traction allows for emergency C0-C1-C2 decoaptation [3,42,43], which clinically translates into an improvement in the neurological state immediately after placement of the Halo-vest. In our series, 8 patients had a spectacular improvement in the neurological state 3 to 6 days after this first stage of care with a significant gain for the mJOA score and Goel grade. In addition, general anesthesia with fiberoptic intubation and installation in prone position were performed safely in a child with complete muscle relaxation [36]. Flexion-extension movements of the cervical spine were well controlled by loosening the halo hinges. Spinal cord decompression was performed by enlargement of the foramen magnum, laminectomy of the posterior arch of C1 and resection of the atlonto-occipital ligament, often adurant to the dura mater [18,36]. The realization of the cranial-C2 fusion with the halo-vest seems satisfactory to us with a successful arthrodesis in all patients and the least risk of perioperative complications. The result of the 2-stage procedure was considered optimal in all 21 cases in our series, as well as that of Tirado-Caballero J et al [3]. Several limitations to the present study need to be addressed. This report represents a retrospective study, single-center series and inconsistent disease type. The

experience of more groups was mandatory to gather enough clinical evidence regarding this issue. Also, further studies were needed to analyze the long-term effects of these craniocervical fusions when the patients become adults.

Conclusion

The 2-stage approach to CCJ anomalies in children with MPS diseases was a safe, inexpensive and effective treatment. There were several advantages to this technique. Mainly, it allows in an emergency situation the cranio-cervical decoaptation and head stabilization in the presence of a neurological deficit. Secondarily, satisfactory fusion of CCJ and good clinical results were obtained in growing children.

Declarations

Funding

No funding.

Availability of data and material

All data is available for the reading committee.

Ethics approval

All data including photos are anonymous.

Summary sentence

Spinal involvement, in particular spinal cord compression at the craniocervical junction, is almost constant in children with MPS. Screening is based on spinocerebral MRI. Two-stage occipito-C2 arthrodesis has proven its effectiveness.

Cranio-cervical decompression associated with non-instrumented occipito-C2 fusion in children with mucopolysaccharidosis: report of twenty-one cases.

Conflicts of interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.xnsj.2022.100183.

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