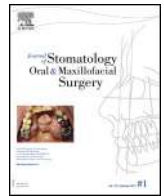




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Case Report

Arthrogryposis multiplex congenita and limitation of mouth opening: Presentation of a case and review of the literature

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ABSTRACT

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Arthrogryposis multiplex congenita (AMC) is a rare entity gathering many diseases, isolated or included in more global syndromes. It affects symmetrically joints of lower limbs, back and maxillofacial area with temporomandibular joint (TMJ) impairment leading to deterioration of oral functions and mouth opening resulting in poor quality of life. Therapeutic strategy varies according to type and severity of impairment and could involve physical therapy and/or surgery. This article reported a case of AMC followed in our unit and highlighting various maxillo-facial impairments, especially restricted mouth opening and its consequences due to fibrotic TMJ and hypertrophic coronoid processes. We described our therapeutic approach based on surgery and postoperative physical therapy. A stable improvement of mouth opening was obtained, and patient's quality of life was increased. We performed a review of the literature and suggested a therapeutic management of mouth opening limitation in AMC.

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1. Introduction

Arthrogryposis multiplex congenita (AMC) is a condition defined as multiple non-progressive and usually symmetrical joint contractures. It can be classified into three main groups depending on which limbs and/or other body parts are involved and on central nervous system dysfunction [1].

Incidence is around 1–3 per 10,000 births. The wrists, shoulders, elbows, hips, knees and ankles are usually affected, with varying degrees of impairment. The oral and maxillofacial region is involved in 25% of patients variably resulting in cleft palate, micrognathia and temporomandibular joint (TMJ) dysfunction with restricted mouth opening, muscle weakness and coronoid hyperplasia. These impairments significantly affect oral quality of life in terms of mastication, swallowing, speech and oral hygiene [2,3].

AMC is a syndromic diagnosis that denotes a large number of diseases, whose exact aetiology, while mainly unknown, is based on neurogenic, myogenic, or environmental factors that limit foetal movements and joint development [4,5]. Standard treatment consists of surgery and physical therapy, especially for orthopedic lesions [5].

Patients with maxillofacial dysfunction, in particular of the TMJ, can have difficulty in eating, and obtaining dental care for the treatment of limited mouth opening. There is no clear consensus on treatment strategy. The main surgical options proposed in the literature are coronoidectomy, TMJ arthroplasty, TMJ replacement sometimes in association with orthognathic procedures. Other surgical approaches (intraoral, extraoral and endoscopic) have to be adapted to the perioperative environment after consultation with anaesthetists. Limited mouth opening, micrognathia, high-arched palate, limited cervical movement or cervical instability, and abnormal orofacial musculature can make direct laryngoscopy or intubation difficult or impossible [6]. Physical therapy should be intensive and long-term in all such cases. We report a case of AMC with limited mouth opening due to coronoid hyperplasia, extra-articular fibrosis and onset of TMJ arthrosis. We present our

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Fig. 1. Limited maximum mouth opening associated with AMC.



Fig. 2. CT scan showing an osteophyte on the lateral side of the left condyle and irregular cortex of the right condyle.



Fig. 4. Osteotomy of the zygomatic arch and transfer of the posterior temporal muscle flap into the posterior part of the recreated TMJ space.

therapeutic approach, discuss the results and give a synthetic review of the literature.

2. Presentation of case

A 38-year-old Caucasian male with a past medical history of AMC presented to the outpatient department of maxillofacial and plastic surgery of the university hospital of Clermont-Ferrand for limited mouth opening. There was no family history of the disorder. He was referred to our department for dental care by his dental practitioner, who was greatly hindered by the impaired mouth opening.

Clinically, the patient had a mouth opening limited to 3 mm and thus precarious oral status including severe parodontopathy with tooth decay and tartaric gingivitis (Fig. 1). He had severe contractures of the wrists, predominantly on the left side, that greatly interfered with his functional activity. He had undergone

surgery on his right wrist several years before. His hips, knees and ankles were affected to such an extent that he could no longer walk and had to use a wheelchair. Panoramic radiography followed by CT scan showed bilateral coronoid hyperplasia and severe extra-articular ankylosis related to fibrotic lesions involving both TMJs. The TMJ spaces were reduced on each side. No severe osteolytic lesions of the mandibular condyle or the mandibular fossa were found. The left condyle was the seat of a small osteophyte on the lateral side and the right condyle had a discreet irregular cortex with loss of regular sphericity (Fig. 2).

It was decided to perform bilateral coronoidectomy and TMJ arthroplasty with temporal muscle interposition but without TMJ replacement under general anaesthesia with nasotracheal intubation. The procedure combined intraoral and extraoral approaches. Disinsertion of the temporal muscle tendon and lower osteotomy

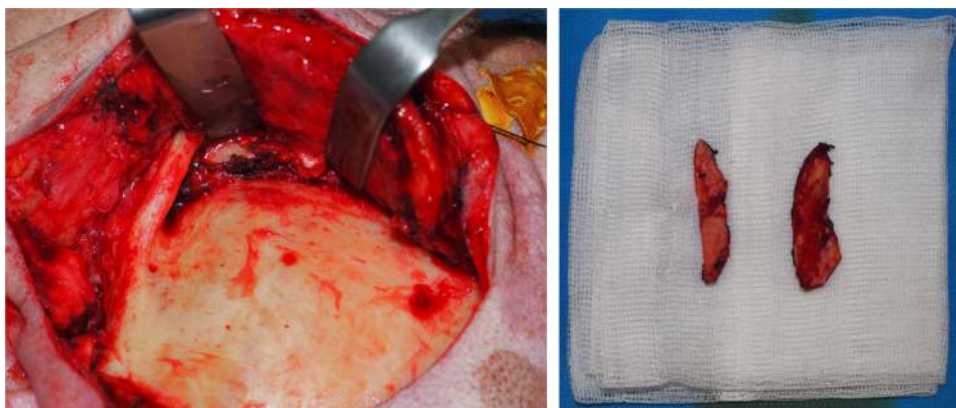


Fig. 3. Left coronoidectomy with an upper extraoral approach. Coronoidectomy fragment.



Fig. 5. 1-year postoperative mouth opening assessed to be 19 mm.

of the coronoid process were performed with an intraoral postero-inferior vestibular approach. The coronoid process fragment removal was achieved by an upper extraoral approach (Fig. 3). A preauricular incision was made as described by Ginestet to allow wide exposure of the TMJ, temporal muscle fossa and zygomatic arch. We decided to free the fibrotic lateral pterygoid and masseter muscles and to perform resection of the uppermost extremity of the condyle followed by contour regularization with a round bur. Osteotomy of the zygomatic arch was then performed, and a posterior temporal muscle flap was transferred, inserted and fixed by three stitches with a 3.0 non-absorbable suture into the posterior part of the recreated TMJ space (Fig. 4). The patient's mouth opening was measured, and the interincisor distance was assessed to be 25 mm passively, which enabled us to perform avulsions of the upper right lateral incisor, upper right third molar, upper left lateral incisor, upper left canine, upper left second premolar, lower left central incisor, lower left first premolar, lower left second premolar, lower right central incisor, lower right canine and lower right first molar.

There were no postoperative complications and the patient left hospital with a prescription for physical therapy that had to be intensive and long-term to maintain improvement of the mouth opening. One week postoperatively, jaw range of motion exercises were started three times a day with use of a TheraBite® system.

At 6 weeks postoperatively, the patient's mouth opening was 17 mm, and 21 mm with manual assistance. He had no pain or other abnormalities. He was recommended to continue intensive physical therapy and rehabilitation of the TMJ up to 1 year postoperatively. The patient continued to use the TheraBite® system, was able to handle utensils, place his fingers in his mouth, brush his teeth, and protrude his tongue, all of which potentiated his recovery.

At 1-year postoperatively, his clinical condition was stable, and his mouth opening was measured to be 19 mm without manual assistance and 24 mm with (Fig. 5), which enabled him to eat solid food again. His speech had improved, and he was able to visit his dental practitioner to consider dental care or at least rehabilitation. He continued four times a day to perform the exercises taught him by the physical therapist.

3. Discussion

Arthrogyposis multiplex congenita is a relatively uncommon diagnosis that encompasses more than 400 conditions that can result in the birth of a child with multiple joint contractures. Because there was little consensus on the management of AMC, guidelines on diagnosis, classification and treatment were drawn

up at the First and Second International Symposia on Arthrogyposis, held in 2007 and 2014, in Birmingham and Saint Petersburg, respectively [7]. No information was given concerning craniomaxillofacial involvement in AMC. One patient in four is affected by AMC, causing severe functional impairments owing to TMJ dysfunction and restricted mouth opening. A literature search was performed in PubMed using "TMJ dysfunction" and "mouth opening" as keywords associated with AMC. The findings are presented in Table 1. One aim of the search was to propose a new and revised treatment algorithm based on the accumulated knowledge of paediatric and orthopedic surgeons [5].

Therapeutic management of TMJ consists in limiting the extent of the mouth opening and its consequences. If nutritional status is satisfactory and/or if dental care can be sufficiently well provided, surgery is not necessary as first-line treatment. However, physical therapy is essential and must be strictly adhered to in the long term [8]. When surgery is indicated, different stepwise approaches can be considered (Fig. 6).

Many reports have advocated bilateral coronoidectomy to increase condylar mobility. Owing to elongation of the coronoid process and to tension of the hypoplastic, fibrotic or atrophic temporalis muscles, the TMJ lacks mobility; however, removal of the coronoid process can easily improve TMJ movement [1,8–10]. Epstein and Wittenberg reported unsuccessful treatment of limited mandibular opening by bilateral coronoidectomy and subsequently by TMJ arthroplasty with release of adhesions, high condylotomy and placement of a Proplast–Teflon disc implant, which improved mandibular function [9].

In our patient, the limitation of mouth opening was extreme, and the condylar unit was morphologically modified. It was therefore decided to perform concomitantly bilateral coronoidectomy and bilateral TMJ capsulotomy and release, associated with condylar remodelling and creation of the joint space by interposition of a posterior temporal muscle flap owing to the risk of exogenous implant expulsion. Nordone and Li reported the case of an Afro-American woman who underwent the same surgery. Postoperative clinical assessment showed temporary improvement, but the patient relapsed because of lack of compliance with physical therapy. A second TMJ arthroplasty was performed with gap arthrolysis maintained by bilateral external distractors (Matthews device arthroplasty) that had been in place for 3 months. At 1-year follow-up, mouth opening and masticatory function were stable. We consider this therapeutic approach to be unnecessarily invasive. In most cases, restricted mouth opening is due to hypertrophic bilateral coronoid processes, masticatory muscle fibrosis and weakness, especially of the temporal and lateral pterygoid muscles, and capsular fibrosis. In some advanced cases of AMC, the condylar unit can be morphologically modified with very small joint spaces. For these patients, condyloplasty with interposition of a temporal muscle flap would be an interesting alternative. Our patient was strictly compliant with postoperative physical therapy and at 1-year follow-up his improvement was maintained. Nordone suggested that it would be prudent to assess patients preoperatively on the basis of their functional and intellectual capacities and also with regard to aesthetic concerns. If the patient is functionally self-sufficient and in good mental health, intensive physical therapy should be encouraged [1]. Patients should be made aware of the necessity of very long-term functional therapy after surgical care. In cases of micrognathia and severe jaw discrepancies related to associated cleft palate or insufficient mandibular growth, orthognathic surgery can be proposed to AMC patients after treatment of restricted mouth opening. Mandibular and maxillary osteotomies should improve both functional masticatory and aesthetic outcomes [3]. Sidebottom reported a case of recurrent TMJ ankylosis treated by TMJ replacement [11]. In our opinion, priority should be given to

Table 1

TMJ impairments and treatments in AMC reported in the literature.

Year	Authors	Type of article	Maxillofacial signs	Paraclinic/imaging	Treatment data	Results
1987	Epstein and Wittenberg	Case report and literature review	Right TMJ pain and restricted mouth opening	Right TMJ: irregularity of glenoid fossa contour Left TMJ: flattening of the anterosuperior aspect of the condyle, shallow glenoid fossa	BC Left high condylectomy and reduction of temporal eminence Teflon-proplast implant (left side) PT	Improved mouth opening (23 mm) Reduction of pain
1988	Hodgson, Weinber, and Consky	Case report	Restricted mouth opening (22 mm) Lateral and protrusive excursions impossible	Left TMJ: grossly deformed with no joint space and Spherically enlarged condylar head Thinned and elongated articular disk	Left condylectomy PT	Improved mouth opening (30 mm)
2001	Jude A. Thomas, Margaret Chiu–Yeh, E. and Steven Moriconi	Case report	Restricted mouth opening (9 mm) Chewing difficulties Dysarthric speech	Menisci badly defined with irregularities Flattened mandibular condyles No bone erosion Severe masticatory muscles weakness	BC BM Lateral pterygoid myotomies Capsular releases PT (TheraBite®)	Improved mouth opening (18 mm) Improvement in speech and jaw function
2003	Lefaiivre and Aitchison	Case report	Restricted mouth opening (6 mm)	No bony fusion between condyles and glenoid fossa Normal joint space Bony extensions between rami and lateral pterygoids muscles	BC Temporal flap inserted in glenoid fossa PT	Mouth opening (25 mm)
2005	Carlos, Contreras, and Cabrera	Case report	Restricted mouth opening Difficulty with feeding and oral care	Severe enlargement of coronoid processes	BC PT Mouth opening device	“Normal” reestablished mouth opening
2007	Kargel, Dimas and Chand	Case report and literature review	Restricted mouth opening: MIO at 25 mm, 15 mm anterior open bite, 10 mm functional mobility Malpositioned dentition within and between each arch Discrepancy of upper and lower jaws	No data available	Orthodontical treatment Le Fort I osteotomy (advancement and posterior impaction) BSSO (3 mm of setback) BC	Improved mouth opening (31 mm) Dental malpositions reduced by orthodontics treatment
2010	Nordone and Li	Case report	Restricted mouth opening (15 mm) Absence of lateral and protrusive movements	Bilateral hypertrophic condylar head and loss of normal joint anatomy Bilateral flattening and loss of cortex of the condylar heads Narrowing of bilateral TMJ space with saucerization of the glenoid fossa Degenerative changes in both TMJs	BC and Brisement force Bilateral open joint arthroplasty + condylectomy + temporalis fascia Bilateral gap arthrotomy + distraction with craniomandibular fixator	MIO at 25 mm Improvement of masticatory function
2010	Puche et al.	Case report	Restricted mouth opening No facial asymmetry	Bilateral coronoid hypertrophy	BC Dynamic physiotherapy	Normal mouth opening at 2 years postoperatively

Table 1 (Continued)

Year	Authors	Type of article	Maxillofacial signs	Paralclinic/imaging	Treatment data	Results
2013	Sidebottom	Short communication	Restricted mouth opening Pain	2008: ankylotic tissue (anteromedially) and dislocation of the prosthesis on the left side 2012: recurrence of ankylosis on the medial side of the joint and reformation of the coronoid process	1995: bilateral TMJ replacement with a Christensen implant then temporary acrylic spacer introduced 2008: bilateral revision replacements + periprothetic fat grafts 2012: repeated arthrotomy, removal of prosthesis, excision of ankylotic tissue, replacement by prosthesis and pack abdominal fat around prosthesis and in the gap PT (TheraBite [®])	Final MIO improved at 26 mm Full normal diet
2015	Balkin, Chen, Oberoi, and Pomerantz	Case report	Restricted mouth opening (5 mm) with only liquid diet Normal masticatory muscles function Incisors in a positive overjet and deep overbite relationship	Enlarged coronoid processes Absence of TMJ ankylosis	BC with AlloDerm insertion into the joint PT (TheraBite [®])	Improved mouth opening (10 mm) Improvement for speech and feeding Persistence of positive overjet and overbite relationship

BC: bilateral coronoidectomies; BM: bilateral meniscectomies; RMO: restricted mouth opening; PT: physical therapy; MOI: maximal interincisival opening.

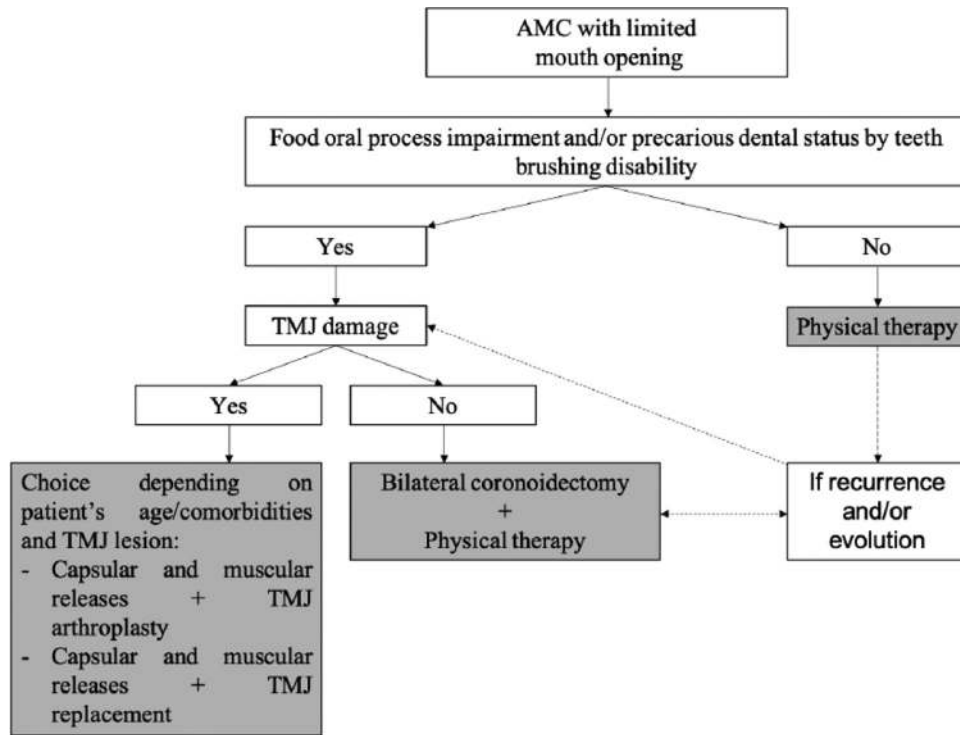


Fig. 6. Therapeutic management of limited mouth opening in AMC.

conservative surgery and, when other therapies have failed or if very severe ankylosis involving bony and articular structures is observed, to TMJ prosthetic replacement.

Limited mouth opening in AMC patients requires rigorous clinical and radiological assessment and needs stepwise functional and surgical care. Long-term physical therapy is essential to avoid relapse.

Disclosure of interest

The authors declare that they have no competing interest.

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