

## Congenital Muscular Torticollis: Study of 30 Cases

Meryem Fettah<sup>1\*</sup>, Mohammed Mimouni<sup>1</sup>, Mohamed Tazi Charki<sup>1</sup>, Hicham Abdellaoui<sup>1</sup>, Karima Atarraf<sup>1</sup>, Moulay abderahmane Afifi<sup>1</sup>

<sup>1</sup>Department of Pediatric Traumatology-Orthopaedics, CHU Hassan II Fez, Morocco. Faculty of Medicine and Pharmacy, University Sidi Mohamed Ben Abdellah Fez, Morocco

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\*Corresponding author: Meryem Fettah

Department of Pediatric Traumatology-Orthopaedics, CHU Hassan II Fez, Morocco. Faculty of Medicine and Pharmacy, University Sidi Mohamed Ben Abdellah Fez, Morocco

### Abstract

### Case Series

Torticollis is a rare condition caused by retraction of the sternocleidomastoid muscle (SCM), most often diagnosed in infants and children. The diagnosis is clinical, with the head and neck assuming an asymmetric spontaneous attitude with a mobility deficiency. Spontaneous evolution is generally reassuring, and it is the fibrous forms that necessitate treatment with home postures and physiotherapy. Surgical treatment is reserved for persistent torticollis after the age of 12 to 18 months. We report a retrospective study of 30 children treated for TMC over a period of 12 years, with ages ranging from 2 to 12 years. All our patients underwent surgical treatment, with a satisfactory overall result; one child was taken back for recurrence. The aim of this study is to analyze the various pathological and therapeutic aspects of this condition.

**Keywords:** Torticollis, Sternocleidomastoid Muscle, Pseudotumor, Physiotherapy, Surgery.

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## INTRODUCTION

Congenital muscular torticollis is the third most common neonatal deformity after hip dysplasia and equinus varus clubfoot. It is defined as a permanent, asymmetrical attitude of the head and neck in reference to the plane of the shoulders. It is due to unilateral retraction of the sternocleidomastoid (SCM) muscle, whose complex anatomy perfectly explains the homolateral tilt, contralateral translation and rotation.

The pathogenesis of congenital torticollis is still a mystery, and several theories have been put forward in the literature, including obstetrical trauma, local perinatal ischemia, venous occlusion, intrauterine malposition, growth disturbance, infectious myositis and compartment syndrome. Diagnosis is straightforward in infants, since the clinical examination is usually sufficient when it reveals retraction of the SCM, whether or not associated with a palpable tumour, which is responsible for the asymmetrical spontaneous posture and the head and neck mobility deficit. The examiner looks for facial asymmetry and associated plagiocephaly, as well as for hip dysplasia, foot abnormalities or scoliosis reflecting excessive intrauterine constraint.

The ideal treatment is still a subject of debate. Some recommend simple monitoring, even though

physiotherapy clearly modifies the natural evolution of these torticollis and allows normalization in more than nine out of ten cases. Surgery is never envisaged before the age of 1 year-18 months, and mainly concerns physiotherapy failures. There are many surgical techniques available, with generally good results, but there is always a risk of recurrence [1–3].

## RESULTS

This is a retrospective study concerning a series of 30 cases of congenital muscular torticollis in children. This work was carried out in the Department of Pediatric Traumatology and Orthopaedics at the Hassan II University Hospital in Fez over a 12-year period from January 2010 to July 2022.

Patient age ranged from 2 to 12 years, with an average of 6 years; 83% of cases were female. The notion of a breech presentation at delivery was found in 30% of cases. For associated anomalies, 2 patients had congenital hip dislocation.

Clinical examination revealed swelling of the SCM muscle in 5 infants, and retraction of the muscle in the rest of the patients (figure 1), with atrophic homolateral muscles, facial asymmetry (figure 2), head turned, flexed, and plagiocephaly in 2 patients.

Standard radiology performed in our patients showed no vertebral malformations or secondary etiologies. Ultrasound showed in 5 infants muscle fibrosis.

Therapeutic management consisted of initial rehabilitation prescribed in 23 patients, i.e. 76% of cases. The evolution was marked by non-improvement in our patients, as physiotherapy was started at a late age. The delay in diagnosis therefore limited the success of physiotherapy.

Two types of surgical technique were used in our patients: 28 children underwent distal tenotomy of the sternal and clavicular heads of the SCM muscle, while 2 patients underwent Z-plasty lengthening of the SCM. Post-operative physiotherapy was indicated in all operated patients for 3 months, after cervical immobilization for 6 weeks.

The mean follow-up period was 3 years, ranging from 8 months to 6 years. The evolution after surgery was favorable in 29 patients, with satisfactory functional and aesthetic results.

A recurrence of congenital muscular torticollis was noted in a single patient, who underwent revision surgery followed by placement of a plaster cast and rehabilitation. The outcome was favourable.



**Figure 1: contraction of the sternocleidomastoid muscle**



**Figure 2: facial asymmetry with shifted shoulders and deviated chin**

## DISCUSSION

The reported incidence of CMT varies from 0.3% to 2% with overall incidence that can be as high as 1 in 250 live births [4–6].

The etiology of CMT and pathophysiology behind SCM impairment is still unknown. Prominent theories behind SCM muscle impairment in CMT include intrauterine crowding, muscle trauma during a difficult delivery, soft-tissue compression leading to compartment syndrome and congenital abnormalities of soft-tissue differentiation within the SCM muscle. Histologic studies of resected surgical specimens have demonstrated edema, degeneration of SCM muscle fibers and fibrosis [7].

The traumatic theory of breech presentation is widely reported, but remains the subject of much controversy [8–10]. In our series, we recorded this presentation in ten observations. A recent study [11], suggests an antenatal origin. Several authors report the frequent association with congenital hip dislocation [8–10], but we found this association in only 2 patients in our series.

The clinical manifestations varies according to age at diagnosis. In newborns, the pathognomonic symptom is an “olive” palpated at the SCM [10, 12]. In most cases, this olive disappears within two to six months, and may be replaced by a muscle retracted [8]. In older children, the most common presentation is a head tilt toward the affected side and the chin pointing to the contralateral side. Plagiocephaly is reported in up to 90% of children with CMT. With continued unilateral weight bearing, the skull base and cranium deform so that the vertex view reveals a parallelogram shaped head. If not treated it may lead to craniofacial growth deformity in adult [13].

Before confirming the diagnosis of CMT, certain secondary torticollis etiologies must be ruled out: postural torticollis, vertebral malformation, malformative syndrome, post-traumatic torticollis, infectious or inflammatory torticollis, or torticollis due to tumour pathology [8]. Cervical ultrasound can be of great help in this respect, and is also of prognostic interest [14]. Radiological investigations (standard X-ray, CT scan, MRI) can be used to study the impact on the spine and eliminate certain differential diagnoses. Ultrasound is the imaging modality of choice for radiographic evaluation of CMT. The normal SCM on ultrasound presents as a hypoechoic mass with echogenic lines, indicating muscle fascicles running throughout its length. The presence of a SCM tumor affects not only the size of the muscle noted on ultrasound, but also its signal intensity. CMT muscles tend to be more hyperechogenic [7].

Progression and prognosis vary from series to series, with spontaneous regression ranging from 70 to

80% [8], all the more so if rehabilitation is initiated early. Unfortunately, in our series, all patients required surgery, as rehabilitation was initiated late [9-12]. Active rehabilitation can be started as early as the 2nd month, the aim being to restore elasticity to the SCM and strengthen contralateral muscles [9-12].

The indication for surgical treatment should not, however, be delayed [15, 16], especially when definitive muscle retraction has set in beyond 18 months, with some authors recommending waiting for olive resorption before any surgery.

Several surgical techniques have been proposed [16], bipolar tenotomy of the SCM is indicated when the retraction is significant in older children. Z-plasty of the SCM offers an aesthetic advantage by preserving the relief of the SCM, the technique we used in 2 patients in our series; and low juxta-clavicular tenotomy via a small surgical approach to both muscle heads, the latter technique we used in almost all patients operated on in our series. The operation is followed by six weeks of postural support in a neck brace.

Apart from postoperative aesthetic discomfort, and very late forms [10-16], the evolution and prognosis of this condition are very satisfactory. Recurrence after surgical treatment is often due to fibrosis regeneration or insufficient resection, as observed in our study.

## CONCLUSION

Congenital muscular torticollis is a rare condition that represents only part of the chapter of torticollis in children. Diagnosis is purely clinical, especially if there is a palpable olive within the SCM, and the multiplication of complementary examinations exploring this "pseudotumor" is not necessary. On the other hand, they are necessary on a case-by-case basis in the search for an etiology, especially if the torticollis is traumatic, febrile or painful.

Prior to any treatment, it is essential to be sure that the patient is not suffering from acquired torticollis, some etiologies of which are serious and constitute therapeutic emergencies.

Congenital muscular torticollis usually evolves favorably, and surgery is only appropriate after the age of 12-18 months, with the aim of correcting the shortness of the SCM before secondary deformities set in. The results are generally good from a functional point of view, but at the cost of a sometimes significant aesthetic ransom.

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