



## **A Rare Case of Neglected Congenital Muscular Torticollis in an Adult and Review of Literature**

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### **Authors' contributions**

*This work was carried out in collaboration between all authors. Authors AM and MAE performed the surgery and assured the follow-up of the patient. Author AG managed the literature searches and wrote the first draft of the manuscript. All authors participated to data analysis and correction. All authors read and approved the final manuscript.*

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**Case Report**

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### **ABSTRACT**

**Aim:** We report a rare case of congenital muscular torticollis in an adult and review most of the different treatment approaches reported by authors.

**Presentation of Case:** A 25 year old woman who underwent an unsuccessful surgery for her congenital torticollis in the childhood, presented to our department for the same problem. We performed a unipolar release of the sternocleidomastoid muscle with a large dissection and a resection of a fibrous part of the clavicular head followed by a muscular rehabilitation. Result was satisfactory with an 18 month follow-up.

**Discussion:** Congenital muscular torticollis is rarely seen in adults. There are almost as many treatment protocols for congenital muscular torticollis as authors, with unequal outcomes. Surgical treatments give the best results in adults. We review most of the treatment protocols described in the literature.

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**Conclusion:** As surgeons, we have to deal with every special situation in the aim of respecting the integrity of healthy tissues and providing the best care for good functional and cosmetic outcome.

**Keywords:** Congenital muscular torticollis; sternocleidomastoid muscle; tenotomy; fibrous band resection.

## 1. INTRODUCTION

Torticollis, from Latin *tortus* (*twisted*) and *collum* (*neck*) was defined by Tubby in 1912 as a deformity, congenital or acquired, characterized by inclination of the head to the shoulder, with torsion of the neck and deviation of the face. [1] Congenital muscular torticollis is declared at birth or in the early infancy, and is connected to shortening and fibrosis of ipsilateral sternocleidomastoid muscle [2]. Its incidence varies between 0.3 and 2%, with a male/female ratio of 3:2 [3]. It is the third most common musculoskeletal congenital condition after dislocation of the hip and clubfoot [1]. Precocious care is recommended and most cases resolve with conservative treatment (physiotherapy) within few months after birth [1,4]. However, rare cases of congenital torticollis in adults can still be seen, especially in poor and developing countries.

We present the case of an adult female living with a congenital muscular torticollis who underwent a successful surgical treatment.

### 1.1 Presentation of Case

A 25 year old woman presented in our hospital suffering from a permanent torticollis without major discomfort. Patient medical history started at birth. Her mother's pregnancy was well attended. Birth was by vaginal delivery. Since then, patient's attitude was characterized by a tilting of the head to the right with a mild rise of the ipsilateral shoulder and the projection of the chin to the left side.

Clinical examination found a shortened, prominent and cord-like right sternocleidomastoid muscle. Compared to the other side, the limitation in rotation of the neck to the left side was about 15° (measured by using an arthrodiagonal protractor). We also noticed the presence of a scar in the right sus-clavicular area which suggests that the patient underwent a previous surgery (Fig. 1). Her parents said that she benefited a surgery in primary childhood during a medical campaign, but they lost the surgical report and don't know what procedure was performed. No other deformity was found. The

clinical presentation was obvious. We didn't perform any paraclinical investigation such as ultrasonography or magnetic resonance imaging because of their difficult access and the priority given to patients with severe conditions and emergencies.

The patient was admitted for a surgical approach. We performed a unipolar release of the right sternocleidomastoid muscle under general anesthesia. We used the same supraclavicular incision as used in the first surgery in order to limit skin damages and to revise the scar. Fibrosis was widely dissected and separated from the healthy muscle (Fig. 2). An excision of the fibrous part of the clavicular head was performed.

A Philadelphia brace was maintained for 10 full days, then partially during a month. Three months of regular muscular rehabilitation including passive and active stretching and physiotherapy (supervised by a physiotherapist) were necessary. The patient was recommended to do active exercises of neck rotation and extension several times a day in front of a mirror. She continued to do so for months after the end of physiotherapy. Outcome after surgery and muscular rehabilitation was highly satisfactory for the patient and her parents. Lee's scoring system (Table 1) was used to evaluate functional and cosmetic discomfort before and after treatment. Improvement was obvious since Lee's score raised from 9 before surgery to 15 and was maintained after an eighteen-month follow up.

Neck rotation and lateral flexion were almost fully recovered without recurrence. There was no clinical evidence of an injury of the accessory nerve. Moderate asymmetry persisted in the neck without any functional significance (Fig. 3).

## 2. DISCUSSION

The diagnosis of congenital muscular torticollis is performed by clinical examination. In birth or soon after, an inclination of the head with the deviation of the chin to the opposite side appear. Contracture of the sternocleidomastoid muscle may be difficult to find in a new born with a short neck. In the neglected cases, a contracture or a

palpable mass may be found in a shortened sternocleidomastoid muscle. The ipsilateral shoulder is often elevated. Asymmetry and skull deformities are more often seen in adult cases.



**Fig. 1. Presurgical view showing a shortening of the right sternocleidomastoid muscle and a sus clavicular scar related to a previous surgery**



**Fig. 2. Surgical view of the clavicular head fibrous band**



**Fig. 3. Postsurgical view**

**Table 1. Lee scoring system for assessment of muscular torticollis [20]**

Points	Function			Cosmesis		
	Facial asymmetry	Neck movement	Head tilt	Scar	Loss of column	Lateral band
3	None	Full	None	Fine	None	None
2	Mild	Limited	Mild	Slight	Slight	Slight
1	Moderate	10°-25°	Moderate	Moderate	Obvious but cosmetically acceptable	Obvious but cosmetically acceptable
0	Severe	>25°	Severe	Unacceptable	Unacceptable	Unacceptable

Ultrasonography is an effective tool of diagnosis. It can help to define the stage of the disease for a better treatment choice [5]. Magnetic resonance imaging can also be useful by showing fibrosis and muscle atrophy [2]. Its use may be limited by a difficult access in developing countries. When indicated, it can be easily performed in adults, but needs a general anesthesia in children.

Sometimes, the patient does not complain of major discomfort for several years until he complains of pain and discomfort in the neck, upper back area or the head, spontaneously or after a trauma [2,6]. In our case, torticollis was well tolerated despite the limitation of neck rotation and lateral flexion.

Some differential diagnosis of congenital muscular torticollis may need a special care or may have a poor prognosis. It is urgent to

identify non-muscular torticollis such as the torticollis related to ocular or hearing deficits. Sandifer's syndrome is characterized by the combination of gastroesophageal reflux, dystonia and torticollis. Neurological torticollis may be connected to cerebral or spine tumors or malformations. Osseous origins of torticollis are in majority related to Klippel-Feil syndrome and rotatory cervical instability [7].

It is reported that prenatal and perinatal circumstances are connected to the appearance of congenital muscular torticollis as: fetal malpositioning in uterus, birth trauma, ischemia of the sternocleidomastoid muscle and compartment syndrome [3,8]. However, the pathogenesis of congenital muscular torticollis remains mysterious. Some recent findings suggest a correlation between some genes expression and the severity of the disease and conclude to the hypothesis of a developmental

disorder of the sternocleidomastoid muscle. This anomaly is characterized by fibrosis, ending up with the shortening of the sternocleidomastoid muscle and all its consequences on the head position [8].

Connections between congenital torticollis and craniofacial abnormalities are well known. Different degrees of abnormalities can be seen. A mild retrusion of ipsilateral forehead and zygoma are often seen. In rare severe cases, deformities involve orbit, nose and mandible. Such cases may need surgical reconstruction [9]. Facial asymmetry cannot be completely corrected after the period of growth potential has passed. It seems that 4 years of age is the limit age after which facial asymmetry becomes difficult to resolve [2]. Seo et al, [10] in a retrospective study including 123 children who underwent surgery for congenital torticollis, suggest that facial asymmetry is progressive, although cranial asymmetry is already determined in children younger than 6 months. In our patient, we didn't notice any clinical facial or cranial deformity.

There is no ideal treatment, but most of the authors suggest an early treatment to avoid deformities [4,9]. Early protocols include passive stretching exercises which can be performed at home when no physiotherapist may be available especially in developing countries. If a concise protocol is properly conducted, results seem to be very encouraging in children before 1 year of age [4]. After 1 year of age, surgery is recommended, and provides its best outcomes before 4 years of age [1]. Some authors recommend delaying the surgical treatment until the patient can comply with postoperative bracing and muscular rehabilitation [2]. Shim and Jang [11] suggest that compliance, rather than age, determines the ideal timing.

Rare neglected cases of congenital muscular torticollis in adults were reported with good outcomes after treatment in most of cases (Table 2). Botox injections were reported to be used as a non surgical approach, with different outcomes [6,12]. An endoscopic surgical approach was used in children and adolescents with good results [13,14]. Liu et al. [15] used radiofrequency therapy in an endoscopic approach with satisfactory results. Open surgical approaches are more often described with different techniques. Open sternocleidomastoid tenotomy was performed in adult patients with

satisfactory outcomes after an average follow-up of 14 years [16]. Bipolar release of the sternocleidomastoid muscle showed satisfactory results in most of the adult patients reported by Omid-Kashani et al. [1] They recommend to release sternal and clavicular heads even if the clavicular head seems trivial. Bipolar release of the sternocleidomastoid with adjunction of z-plasty was proposed in children by Ferkel et al. [17] in 1983 and recently in adults by Patwardhan et al. [18] with good results. According to the authors, a bipolar release without z-lengthening may lead to the loss of the normal neck contour and affect the cosmetic appearance. Omid-Kashani et al. [1] recommend the z-plasty in all neglected old cases. Recently, Pombo Castro et al. [2] reported a case of congenital muscular torticollis successfully managed by a bipolar release of the sternocleidomastoid using the harmonic scalpel. They claim that compared to classic electric scalpel, the harmonic scalpel provides less bleeding and better post operative comfort. Lee et al. [3] reported a series of 20 children and youths under 20 years of age who benefited a complete release and resection of the fibrous band on the sternocleidomastoid band. They recommend this technique for its good functional and cosmetic results. Amemiya et al. [19] comparing unipolar and bipolar release noticed that 3 from the 5 patients (children and youths) who had a unipolar release needed a second surgery for recurrence. Recurrences are more often described in children and youths than in adults because of a larger number of reports and also because of their ability to grow up. Compliance to post-operative treatment is also a factor of non recurrence for adults.

According to most of the different authors, there was no or almost no improvement in skeletal deformities after any of the different procedures used in adults.

In our case, we performed a unipolar release of the sternocleidomastoid muscle associated with a dissection of the fibrosis which was largely separated from the healthy muscle and a resection of the fibrous part of the clavicular head. We had to manage with the fact that the patient underwent a non documented previous surgery in childhood. We tried to avoid damages of the healthy tissues and nerves (accessory nerve). Outcome was very encouraging.

**Table 2. Different studies and their outcomes**

Author	Number of patients	Sex		Mean age (extremes)	Treatment	Results	Follow-up
		Male	Female				
Bouchard et al. 2010 [6]	3 adults	0	3	30 yo [15 - 39]	Botulinum Toxin injection	Moderate or good results	Needing reinjections to maintain benefit
Burnstein and Cohen 1998 [13]	12 children	7	5	10,5 month [4 - 18]	Endoscopic release of the SCM under general anesthesia	Good to excellent results	14 months [6 - 24]
	1 adolescent	0	1	14 yo	then 4 to 6 months of physiotherapy	One per-operative complication: transection of a branch of the external jugular vein in 1 case	
Tang et al. 2010 [14]	45 children (4 of them have had a previous open surgery for the same problem)	26	19	4,8 yo [0,5 - 14]	Endoscopic release of the SCM under general anesthesia then 4 to 6 weeks of physiotherapy And a neck collar for 1 month for patients older than 3 yo	Excellent results in 88,1% Good results in 9,5% Fair results in 2,4%	18 months [6 - 36]
Liu et al. 2011 [15]	26 adults	11	15	22 yo	Radiofrequency carbonation with local anesthesia under arthroscope	19 excellent outcomes 7 fair outcomes	25 months [15 - 71]
Ippolito and Tudisco 1986 [16]	8 adults			26 yo [20 - 37]	Open tenotomy of the SCM	satisfactory results	14 years
Omidi-Kashani et al. 2008 [1]	18 adults (4 lost during follow-up)	10	4	21,9 yo [18 - 32]	Bipolar release of the SCM Then the use of a Torticollis brace during 3 months and physiotherapy	7 excellent results 5 good results 2 recurrences	2,5 years [1 - 5]
Ferkel et al. 1983 [17]	34 children	12	22	28 months [9 weeks - 7,5 yo]	14 patients had no surgery: exercices alone in 13 cases and cast for 2 weeks and then exercices in 1 patient	86% of excellent or good results	6,5 years for the 13 patients treated by exercices alone and 10,5 years for the surgical patients No reoperation needed
					10 patients had an open tenotomy of the mastoid and clavicular attachments, release of the cervical fascia and z-	92% of excellent or good results	

					plasty of the sternal attachment of the SCM 10 patients had other procedures: 5 patients had a unipolar tenotomy only, and 5 patients had a bipolar tenotomy, NB: 1 patient had 3 procedures and 3 patients had 2 operations each	20% of good results 70 % of fair results 10% of poor results	4 patients needed reoperation
Patwardhan et al. 2011[18]	12 adults			24 yo [17 - 31]	Bipolar release of the SCM and Z-lengthening Then the use a neck collar for 3 weeks And an active assisted exercices for 6 weeks	6 patients had excellent result 2 patients had good results 4 patients had fair results	minimum of 2 years
Pombo-Castro et al. 2014 [2]	1 adult	0	1	35 yo	Release of the SCM from its insertions in the mastoid process, clavicle and sternum and resection of a segment of the muscle using the Harmonic scalpel Then immobilization by a brace and passive and active stretching	Satisfactory result	4 years
Lee et al. 2009 [3]	20 children and youths	13	7	47,6 months [9 - 240]	Selective fibrous band release and resection Physiotherapie for 3 - 4 months 8h/day neck collar for 3 months	Good fonctionnal and cosmetic results	

We believe that the association of a unipolar release and the resection of a part of the fibrous band of the sternocleidomastoid muscle presents a lower risk of recurrence in adults. Till now, our patient is regularly seen and the result is maintained satisfactory. There are no signs of recurrence 18 months after surgery. Eventually, we propose a lipofilling to balance the aesthetic appearance of the neck contour.

### 3. CONCLUSION

Even if congenital muscular torticollis is frequently seen in early infancy and resolve in most cases after a medical approach, some neglected cases can be rarely seen in adults and need surgical treatment. There are almost as many treatment protocols for congenital muscular torticollis as authors, with unequal outcomes. As surgeons, we have to deal with every special situation in the aim of respecting the integrity of healthy tissues and providing the best care for good functional and cosmetic outcome.

### CONSENT

All authors declare that written informed consent was obtained from the patient for publication of this case report and accompanying images.

### ETHICAL APPROVAL

It is not applicable.

### COMPETING INTERESTS

Authors have declared that no competing interests exist.

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