



## Surgical treatment of congenital pseudarthrosis of the clavicle : A report on 17 cases

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**Congenital pseudarthrosis of the clavicle (CPC) is a rare malformation of uncertain aetiopathogenesis, usually unilateral. Physical examination reveals swelling over the midportion of the clavicle, often asymptomatic ; the diagnosis is confirmed by radiology. Treatment is controversial : for many authors the surgical indications are the presence of symptoms, functional impairment or cosmetic deformities. We present a retrospective analysis of 17 children with CPC treated in our institutions : 9 were treated with plate (P) and 8 with Kirschner wire (KW) fixation ; a bone graft was used in 12 cases only. Five patients (4 P and 1 KW) needed a second surgical procedure.**

**The surgical treatment led to a very good result in 7 cases, good in 4 cases, fair in 3 cases and poor in 3 other cases.**

**We recommend early treatment of all patients with CPC with resection of the pseudarthrosis, autologous iliac bone grafting and internal fixation with Kirschner wires.**

**Keywords :** pseudarthrosis ; congenital ; clavicle ; treatment ; review.

### INTRODUCTION

Congenital pseudarthrosis of the clavicle (CPC) is a rare malformation of the shoulder girdle. The aetiopathogenesis is still uncertain. The lesion is usually unilateral : it is localised in the middle part of the clavicle and it mostly affects the right side.

Physical examination reveals swelling over the clavicle ; often the patient is asymptomatic and has no functional limitations ; the diagnosis is confirmed by radiology.

In 1910 Fitzwilliam distinguished this pathology from posttraumatic pseudarthrosis, cleidocranial dysostosis and neurofibromatosis. Due to its frequency and similarity with CPC, differential diagnosis should be made with obstetrical fracture of the clavicle.

Treatment is controversial : for many authors the surgical indications are presence of symptoms, functional impairment and cosmetic deformities.

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Table I. — Patient features

Patient	Sex	Age at Diagnosis	Pain	Function	Swelling	Family History	Age at Surgery	Surgery				
								Resection	Internal Fixation	Graft	Complications	Revision Surgery
1	F	1 y	No	Good	Slight	No	4 y 6 m	Yes	Plate	Yes	Breakage (4 d)	Plate
2	F	Birth	No	Good	Serious	No	7 y 1 m	Yes	Plate	Yes	No	Removal 9 y
3	M	Birth	No	Good	Slight	No	7 y 6 m	Yes	Plate	No	No	Removal 11 y
4	F	3 y	No	Good	Moderate	No	4 y 4 m	Yes	Plate	Yes	No	Removal 6 y
5	F	1 y	No	Good	Serious	No	7 y 4 m	Yes	Plate	No	No	No
6	M	Birth	Yes	Good	Moderate	No	6 y 10 m	Yes	Plate	No	Breakage (3 m)	Plate And Graft
7	F	Birth	No	Good	Slight	No	4 y 10 m	Yes	Plate	Yes	Mobilisation	Plate
8	F	Birth	No	Good	Moderate	No	4 y 7 m	No	Plate	No	Mobilisation	K-Wire
9	M	Birth	No	Good	Serious	No	4 y 10 m	Yes	Plate	Yes	No	No
10	M	Birth	No	Good	Moderate	No	5 y 1 m	Yes	K-Wire	Yes	No	Removal 3 m
11	M	4 y	Yes	Good	Moderate	No	7 y 7 m	Yes	K-Wire	Yes	No	Removal 2 m
12	M	17 m	No	Good	Slight	No	7 y 1 m	Yes	K-Wire	Yes	No	Removal 3 m
13	M	2 y	No	Good	Moderate	Yes	6 y 7 m	Yes	K-Wire	Yes	Infection	External Fixation 45 d
14	F	3 m	No	Quite Good	Serious	No	6 y 9 m	Yes	K-Wire	Yes	No	Removal 3 m
15	F	2 y	No	Quite Good	Serious	No	5 y 4 m	Yes	K-Wire	No	No	Removal 3 m
16	F	Birth	No	Good	Moderate	No	5 y 3 m	Yes	K-Wire	Yes	No	Removal 2 m
17	M	3 y	Yes	Quite Good	Slight	No	7 y 6 m	Yes	K-Wire	Yes	No	Removal 3 m

Legend : (M) Male, (F) Female, (y) Year, (m) Month, (d) Day.

The purpose of this study is to report our experience (17 cases) and to review the existing literature in order to identify the most appropriate indications and surgical technique for treatment.

## MATERIALS AND METHODS

A retrospective analysis was performed on 17 children treated between 1994 and 2003 at the Hôpital des Enfants Malades-Necker of the Université "René Descartes" in Paris and the Dipartimento di Scienze dell'Apparato Locomotore dell'Università "La Sapienza" in Roma (table I).

Our study involved 9 girls and 8 boys ; a family background was noted in one case. Cranial dysostosis, neurofibromatosis and post-traumatic pseudarthrosis were ruled out in all cases.

The lesion involved the right side in all cases, and all patients presented a bump over the middle third of the

clavicle. The pseudarthrosis was detected at birth in 8 cases and when the children were 2 years old (maximum) in 6 cases ; in 2 cases the anomaly was detected fortuitously during a radiographic examination and in one case it was detected during an examination for torticollis. The deformation of the clavicle was detected by persons close to the children in 12 cases, while in 5 cases it was detected by the attending physician or the paediatrician.

Clinical examination showed a swelling in 12 cases while in 5 children the deformation was not visible, but was noted on palpation. Discomfort on shoulder mobilisation was detected in three cases and pain in another three ; vascular or nervous problems were noted in no instance.

Initial radiographs showed discontinuity in the mid-portion of the clavicle with an interfragmentary gap : the bone ends were hypertrophic or thinned ; the sternal fragment was always longer than the acromial fragment, which it overrode.



*Fig. 1.* — Plate breakage



*Fig. 2.* — Plate displacement (fixation failure)

Surgical treatment was performed in 9 cases for cosmetic reasons or upon parental request, in 4 cases due to a defect in the development of the shoulder girdle and 4 times in order to prevent the possibility of further complications. The average age at surgery was 6 years and 4 months (range : 4 years and 4 months to 7 years and 6 months). The procedure involved in all cases excision of the pseudarthrosis tissue and fixation (in 9 cases with a plate and in 8 with Kirschner wires); an autologous corticocancellous bone graft from the ilium was used in 12 cases. Following the surgery, the upper limb was immobilised with a Mayo clinic or Dujarier type of bandage for 45 days on average. In the cases treated with Kirschner wires, the internal fixation was removed after 2 months on average.

## RESULTS

Bone healing was achieved after initial surgery in 12 cases and following revision surgery in 5 cases. Surgical failure occurred in 4 cases following plate fixation, with plate breakage in two cases (fig 1) and fixation failure in the other two (fig 2), and in one case following fixation with Kirschner wires, with infection (fig 3). All patients were reevaluated after an average follow-up of 4 years and 1 month (range : 6 months to 10 years).

At each visit we performed a clinical assessment of cosmesis, symptoms, and function, and a radiological evaluation of bone consolidation (table II).

As regards cosmesis, we took into account the scar and surface appearance of the clavicle. The



*Fig. 3.* — Infected non-union following K-wire fixation

scar was satisfactory (scarcely visible) in 12 cases, visible (hypertrophic) in 3 cases (1 P and 2 KW) and unsightly (cheloid) in 2 cases (2 P). The surface appearance of the clavicle was comparable on both sides in 12 cases, whereas in the remaining 5 cases the shoulder girdles were asymmetric (P 4 and KW 1).

Two previously asymptomatic patients complained of pain (2 P).

Function was unrestricted in all cases, except in one patient in whom we noted decreased muscle strength in the upper limb, probably due to late treatment of the lesion.

We assessed the length of the clavicle radiographically : it was similar to the normal side side

Table II. — Results

Patient	Follow-up						
	Scar	Clavicle Profile	Pain	Function	Rx Length	Results	Duration
1	Satisfactory	Asymmetrical	Yes	Good	-	Insufficient	6 y
2	Satisfactory	Symmetrical	No	Good	=	Good	18 m
3	Satisfactory	Asymmetrical	No	Good	=	Good	2 y
4	Satisfactory	Symmetrical	No	Good	=	Good	3 y
5	Satisfactory	Symmetrical	No	Good	=	Good	10 y
6	Antiesthetic	Asymmetrical	No	Good	-	Insufficient	4 y
7	Antiesthetic	Asymmetrical	Yes	Good	—	Insufficient	7 y
8	Visible	Asymmetrical	No	Good	-	Sufficient	5 y
9	Satisfactory	Symmetrical	No	Good	=	Sufficient	6 y
10	Satisfactory	Symmetrical	No	Good	=	Good	4 y
11	Satisfactory	Symmetrical	No	Good	=	Good	1 y
12	Satisfactory	Symmetrical	No	Good	=	Good	6 m
13	Visible	Asymmetrical	No	Good	-	Sufficient	2 y
14	Satisfactory	Symmetrical	No	Good	=	Good	4 y
15	Satisfactory	Symmetrical	No	Good	=	Good	7 y
16	Satisfactory	Symmetrical	No	Good	=	Good	4 y
17	Visible	Symmetrical	No	Good	=	Good	3 y

Legend : (y) Year, (m) Month, (=) Same length, - Shortening of 0.5 – 1 cm, — Shortening of 1.5 cm.

in 12 cases ; in 4 cases, it was 0.5-1 cm shorter (P 3 and KW 1) and in one case it was 1.5 cm shorter (P).

We considered as negative features the presence of an unsightly scar, pain, functional impairment, shoulder girdle asymmetry due to shortening of the clavicle. In children in which these features were not present, the overall result was rated good ; in the presence of one of these features, it was rated fair and in children with 2 or more features present, it was rated poor. Eleven patients (P 4 and KW 7) achieved a good result, 3 patients (P 2 and KW 1) a fair result and 3 patients (P 3) a poor result.

## DISCUSSION

Congenital pseudarthrosis of the clavicle (CPC) is a rare malformation. Up to now, about 200 cases have been reported in literature ; the largest series were described by Gibson and Carroll (8) (27 cases) and Cadilhac *et al* (5) (25 cases). Males and females

are equally affected. The lesion is normally unilateral and mainly occurs on the right side ; the few cases affecting the left side are usually associated with dextrocardia (8). Rare cases with bilateral involvement have also been described (14,24,27).

Associated malformations reported in the literature are : congenital elevation of the scapula (10), boomerang-shaped scapula (23), bilateral hip dysplasia, interventricular communication (37) and dextrocardia (8).

There is still debate regarding the pathogenesis : the clavicle is the first segment of the foetal skeleton to undergo ossification ; several studies (38) have shown that from the 7<sup>th</sup> week of intrauterine life, two centers of primary ossification are present at the two extremities of the clavicle, which unite in the following weeks due to the process of calcification. Alldred (2) suggested that if these centers of ossification failed to unite, thus developing as two separate segments, CPC could result (not a defect of consolidation but a defect of fusion). This

hypothesis was reappraised and confirmed by studies carried out in 1995 by Hirata *et al* (12) who histologically analysed the tissues of the pseudarthrotic stumps : he found hyaline cartilage to be present on both edges of the clavicular fragments, like the epiphyseal cartilage of other segments in growing age. A possible defect of fusion during intrauterine life was explained by Lloyd-Roberts *et al* (18) in 1970 according to a simple anatomical consideration, namely that the subclavian artery, which runs posterior and inferior to the midportion of the clavicle, has a higher origin and course on the right side and could therefore influence ossification of the clavicle owing to its pulsations. This interesting pathogenic theory seems to be supported by the fact that in about 95% of cases the pathology affects the right side and that the few left-sided cases are in subjects with dextrocardia (in which case the left subclavian artery has a higher origin and course). Familial occurrence of this anomaly has been documented but the pattern of genetic transmission remains unclear : Fernández-Cuesta *et al* (6) reported a case of congenital pseudarthrosis of the clavicle in twins and Price (25) described 2 cases involving a father and daughter.

The deformity is only rarely identified at birth, usually in the first months of life. It generally presents as a subcutaneous swelling over the middle third of the clavicle, which tends to increase in size as the child grows ; in typical cases it is not painful (with some rare exceptions (28)) and does not limit the range of motion of the upper limb. In this context the problem can be considered in terms of cosmesis. During adulthood, pain, functional impairment and compression syndromes of the neuro-vascular structures may develop (3,7,11,21).

Several different surgical procedures have been advised consisting of removal of the pseudarthrotic tissue, bone grafting and alignment of the stumps with a plate or K wires (9,17,20,23,26) but there is no full agreement whether patients should be treated while they are still asymptomatic and functionally fit (36). Certain authors opted for treatment abstention. Wall (38), Kite (15) and, more recently, Shalom *et al* (30) took into account the risks of surgery (failures, cosmesis damage, complications) and found it preferable not to perform any surgery

if the problem is merely cosmetic. Toledo and MacEwen (33), on the other hand, advocated a conservative treatment after being confronted with a motor and sensory paralysis of the brachial plexus following an osteosynthesis with K wires. For some authors the indication for surgery was neuro-vascular (3,7,8,11,19,21,29,31,32,35). In the literature, the indication for surgery has been cosmesis in the majority of cases (4,8,17,23). Grogan *et al* (9), Shoenecker *et al* (29) and Ahmadi and Steel (1) were led to perform the surgery upon request from the patients or the parents, for cosmesis reasons added to the discomfort felt by the patient. Hirata *et al* (12), Jinkins (13) and Kohler *et al* (16) performed surgery on all children in order to avoid possible consequences on shoulder development while Schnall *et al* (28) and Ullot *et al* (34) operated only on children who had localised pain at the pseudarthrosis site. Marmor (22) based his surgical indication in adults on the onset of pain, and Gibson and Carroll (8) recommended treatment of all adults.

## CONCLUSIONS

Based on our experience and the literature review, the most effective surgical procedure involves resection of the pseudarthrosis, iliac bone grafting and internal fixation with Kirschner wires. We suggest early treatment in all cases, even though in the initial stage CPC appears to be a "simple" cosmetic defect, as in the course of time the swelling tends to enlarge, thus causing the onset of symptoms and functional impairment. Finally, surgery performed in children is less aggressive than in adults.

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