



## Congenital pseudarthrosis of the clavicle : The role of CT-scanning

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Congenital pseudarthrosis of the clavicle is a rare entity. It presents on the right side in 90% of the patients, and bilaterally in up to 10%. The authors report the case of a 4-year-old boy who presented with a painful deformity over his right mid-clavicular area. Plain radiographs were inconclusive, although the opposite is true in most cases. A computed tomography (CT) 3-D reconstruction showed a pseudarthrosis of the clavicle, and excluded a neoplastic, infective or traumatic origin. Treatment involved excision of the pseudarthrosis, internal fixation with a contoured reconstruction locking plate, and bone grafting. The authors prefer operative treatment, but this is not universally accepted.

**Keywords :** congenital pseudarthrosis ; clavicle ; computed tomography ; internal fixation.

### INTRODUCTION

In 1977 Ahmadi and Haldeman (1) found only 102 cases of congenital pseudarthrosis of the clavicle in the world literature. The aetiology is unknown. However, a positive family history has been described. Most authors agree that if this condition is indeed transmitted genetically, it is probably a recessive type. Wall (10) feels that the defect might not be a result of failure of fusion of the two primary ossification centers in the desmal phase of ossification, but rather a result of atresia of blood vessels when the original desmal bone mass is resorbed during the phase of endochondral ossification.

### CASE REPORT

A 4-year-old boy presented to the paediatric orthopaedic clinic with an intermittently painful deformity over his right clavicle (fig 1). First noticed by the parents, soon after birth, it had been diagnosed on plain radiographs at age 4, as a fractured clavicle. Since one year it had been increasing in size and was causing the patient discomfort and distress due to the cosmetic appearance. There was no history of trauma, and the patient was otherwise medically normal. Clinical examination revealed a firm, mildly tender, ugly deformity over the middle third of the right clavicle. There was a full range of active and passive movement in the right shoulder joint without other positive clinical findings.

Plain radiographs (fig 2) did not show the classical features of a congenital pseudarthrosis and were described as a possible old fracture with callus formation, however taking into account the

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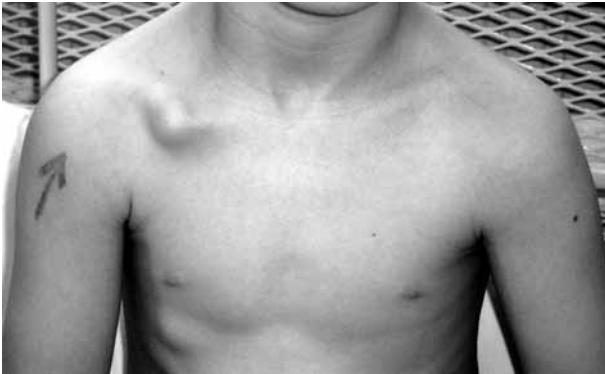
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*Fig. 1.* — Clinical deformity seen over the middle third of the right clavicle, showing significant cosmetic abnormality.



*Fig. 2.* — AP radiograph of the right clavicle, showing a swelling over the middle third of the clavicle, without the classical features of a pseudarthrosis.



*Fig. 3.* — CT-3D reconstruction clearly demonstrating the pseudarthrosis.



*Fig. 4.* — Postoperative AP radiograph showing fixation of the clavicle with a reconstruction locking plate and unicortical screws.

differential diagnosis of Ewing's sarcoma, Caffey's disease and osteomyelitis. A Computed Tomography (CT) scan with 3-D reconstruction (fig 3) clearly demonstrated a pseudarthrosis of the middle third of the right clavicle, thereby confirming the clinical diagnosis and excluding a suspicious cause for the clinical and radiological appearances.

As the patient was symptomatic, and the cosmetic deformity significant, the pseudarthrosis was excised, and internal fixation was performed (fig 4), using an AO reconstruction locking plate, with a bi-cortical bone graft obtained from the iliac crest, and unicortical screws. The postoperative period was unremarkable. After 3 months there was bony union and a good cosmetic appearance. The

patient and his parents were pleased with the outcome.

## DISCUSSION

Congenital pseudarthrosis of the clavicle is a rare condition. The true incidence is unknown. Ninety percent occur on the right side, and up to 10% are reported as being bilateral (1, 3). The cosmetic deformity is usually progressive, but pain is often not associated with the lump on the clavicle (10).

Plain radiographs are said to be distinctive. Typical features are involvement of the middle third of the clavicle, definite separation of the bone

ends, lack of reactive callus, and bulbous appearance of the lateral segment at the pseudarthrosis without tapering of the bone ends (2, 8).

The differential diagnosis usually consists of posttraumatic pseudarthrosis and cleidocranial dysostosis (10). The latter affects other bones as well: cranium with wide fontanelles and wide suture lines, small facial bones and crowded teeth; discrete skeletal anomalies in the hands and feet; defective ossification of the pelvis; neural arch defects (1, 4). On the other hand, postpartum fractures hardly ever lead to a pseudarthrosis. Posttraumatic pseudarthrosis usually shows exuberant callus formation and is associated with the clinical features of pain and tenderness on examination (2).

In this case the characteristic radiological features described above were not present as the pseudarthrosis was hidden behind the apex of the deformity. The increasing size over the last year, the pain and the radiological features also made a neoplastic or infective cause a possibility. The CT-scans and particularly the 3-D reconstruction images were of vital importance in reaching a diagnosis and formulating a treatment plan in this particular case. They allowed to clearly visualise the pseudarthrosis in a 3-dimensional manner. Plain radiographs are only able to give a 2-dimensional representation of the problem, which in this case proved insufficient to give an accurate diagnosis.

Treatment of this condition varies. If the patient is asymptomatic then conservative treatment has been advocated, as it is felt to be a benign condition (1, 4). Conversely it has been reported that, if treated conservatively, the deformity can increase. This may lead to instability of the shoulder girdle and to neuro-vascular complications. In the authors' opinion operative treatment is therefore recommended, usually before school age (5, 7). However, this approach is not universally recommended (1, 10).

Operative intervention involves resection of the pseudarthrosis, with or without bone graft interposition, and internal fixation. If no internal fixation is used, techniques have included maintaining a

periosteal sleeve around the clavicle to help approximate the bone ends (6). Different types of internal fixation have been suggested in the literature. Steinmann pins and Kirschner wires (9) have led to serious complications, such as migration, pin tract infection and non-union. Recent reports (5, 8) plead for reconstruction with a plate and bone grafts.

The authors highlight the role of the reconstruction locking plate for the fixation of clavicle and bone graft after excision of the pseudarthrosis. This plate can be moulded to follow the natural sigmoid shape of the clavicle prior to insertion, allowing to recreate the natural contour of this bone. The use of unicortical screws further reduces the risk of injury to the important neurovascular structures which run inferiorly and posteriorly to the clavicle.

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