

## Congenital Pseudarthrosis of the Clavicle

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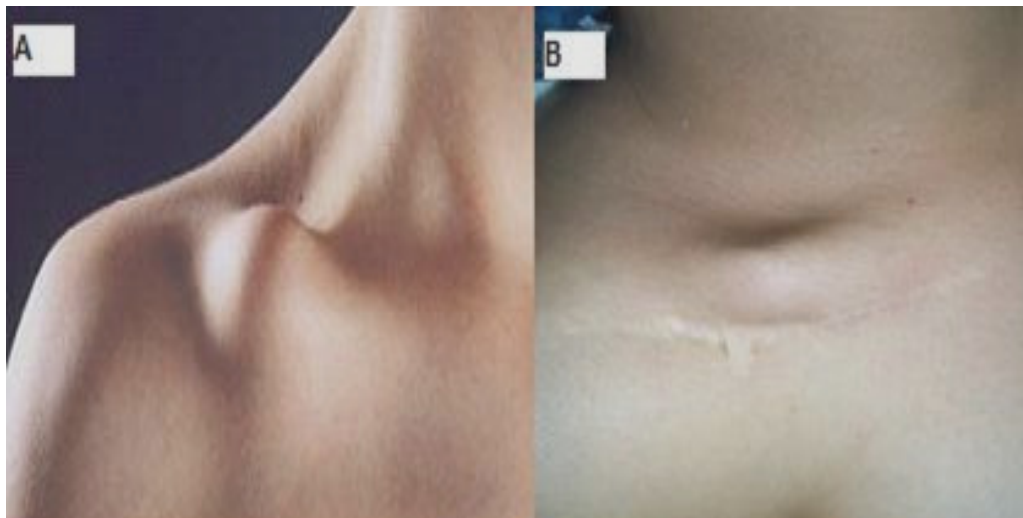
**Congenital pseudarthrosis of the clavicle is a rare condition and till date only about 200 cases have been reported in literature. Once such rare case of congenital pseudoarthrosis affecting right clavicle of a girl is presented .**

### Introduction

Congenital pseudarthrosis of the clavicle is a rare condition and after being reported for the first time in 1910, only about 200 cases have been reported in literature in last hundred years. The condition is predominantly right sided and etiology is still unclear. Imaging plays a vital role in diagnosis and subsequent management. Surgical management comprises of excision of pseudarthrosis with internal/external fixation of bony ends with or without bone grafting. Due to rarity of this condition, a case of a girl reporting with congenital pseudarthrosis of right clavicle is presented.

### Case presentation

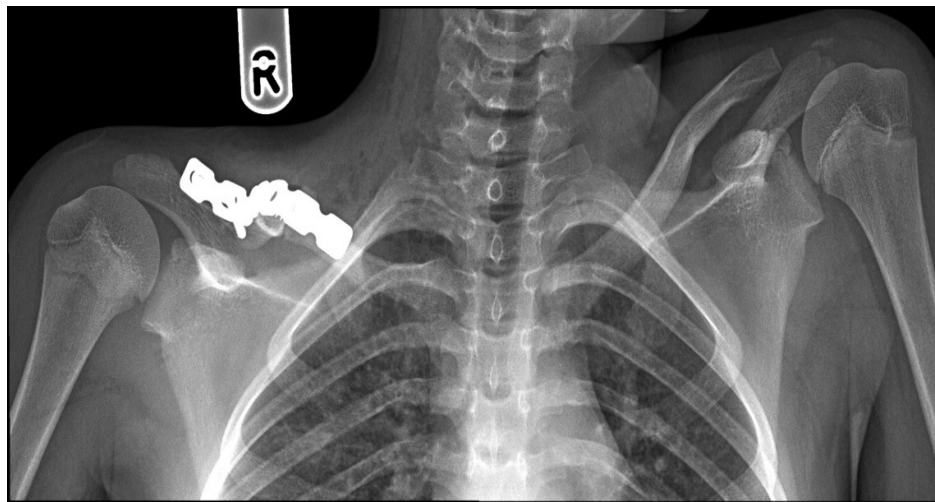
A 12 years old girl, first in birth order was brought by parents with a unsightly prominence over the right clavicular area. The swelling had been noticed by parents since her birth and had been growing in size gradually as the patient grew up with age. The girl was first in birth order and had no other medical, surgical or family history of significance. There was no history of a difficult delivery or injury. On examination, the patient was of average built and had a non tender bony lump lateral to the middle of right clavicle. The skin over the lump had no abnormal features. The size of the lump increased with abduction of the right shoulder. The ranges of motions of right shoulder were normal. Plain images showed characteristic pattern of congenital pseudarthrosis of right clavicle with a defect in the diaphysis and anterior and superior tilting of the sternal half and a smaller acromial portion with bulbous ends. The CTscans with the 3-D reconstruction images showed the bipartite right clavicle with no evidence of developing callus or soft tissue mass and confirmed the diagnosis.



**Figure 1.** Preoperative (A) and two years postoperative follow-up (B) images of congenital pseudarthrosis of right clavicle.



**Figure 2 .** Plain image (A) and 3- D reconstruction CT scans ( B, C, D) showing features of congenital pseudarthrosis of right clavicle.



**Figure 3.** Follow up plain images at 6 months after surgery of congenital pseudarthrosis of right clavicle.

The patient was transferred to the care of pediatric orthopedic surgeon and underwent operative intervention involving resection of the pseudarthrosis, with iliac bone graft interposition, and internal fixation. The patient was seen after 2 years for a different reason. She was satisfied with the outcome of clavicular surgery. She had full range of movement of the right shoulder and arm, and was pain-free. There was minimal residual prominence over the lateral half of right clavicle and a subclavicular scar which was widened over a length of 3 cm laterally. She was offered the option of scar revision under local anesthesia.

## Discussion

Congenital pseudarthrosis of the clavicle is a rare disorder of the shoulder girdle. The condition was initially described by Fitzwilliams<sup>1</sup> as a distinct variant of cleidocranial dysostosis in 1910 and since then with only approximately 200 individual cases having been reported in the world literature to date<sup>2</sup>. It presents predominantly on the right side (about 90%) of the patients<sup>2,3</sup>. The left sided involvement usually occurs with dextrocardia and situs inversus<sup>4</sup>. Bilateral cases can occur but are rare and are typically associated with genetic syndromes<sup>5</sup>. The exact etiology remains unknown but it is believed that the lesion is caused by pressure exerted upon the developing clavicle by the pulsating subclavian artery. The right subclavian artery is normally situated at a higher level, which explains the predominant right clavicular involvement<sup>6</sup>. Another theory suggests the condition to be due to the separation of the two primary ossification centers<sup>7</sup>. Bilateral involvement is related to an abnormally high subclavian artery on both sides, caused by cervical ribs or high vertically oriented upper ribs. Some cases occurring in members of the same family have been reported in literature, leading to theories that it may be an inherited disease with an autosomal dominant transmission<sup>8</sup>.

Congenital pseudarthrosis of the clavicle usually presents as a painless prominence lateral to the middle of the clavicle at birth or in early neonatal life. The ends of the clavicular segments are enlarged at the site of the pseudarthrosis and there is some degree of motion between them. The deformity tends to become more obvious and unsightly as the child grows with age. The skin above the prominence may become atrophic. The vertical border of the scapula on the affected side looks asymmetrical and prominent, while the affected shoulder is lower. The deformity becomes greater when the patient abducts the arm. The condition is generally painless but mild to moderate pain may be present around the shoulder girdle and upper arm in some cases. A few patients may also develop functional problems of the upper limb of affected side like weakness, limited shoulder abduction or features of thoracic outlet obstruction<sup>9,10</sup>.

The differential diagnoses of congenital pseudarthrosis of the clavicle include obstetric fracture, post-traumatic non-union, cleidocranial dysostosis, or neurofibromatosis<sup>11</sup>. An obstetric fracture of the clavicle is suggested by history of a difficult delivery, pseudoparalysis of the arm with no voluntary limb movement and pain on passive movement, or when a massive callus is seen radiographically. Traumatic nonunion can be differentiated by plain radiographs as in the non union the bone ends seem attenuated. In cleidocranial dysostosis there are other skeletal abnormalities such as skull deformities, small size of the facial bones, scoliosis and deficiencies of the pelvis. In neurofibromatosis, the bony ends are attenuated and other characteristic stigmata are present which include axillary / inguinal freckling and cafe-au-lait spots on the skin. In this regard, clavicular pseudoarthrosis is different from tibial pseudoarthrosis. In tibial pseudoarthrosis, 50% of cases are associated with neurofibromatosis and in such cases there may be a lytic lesion seen within the bone.

The deformity is evaluated with imaging which generally include plain radiographs and CT scan. Anterior, posterior, and apical lordotic views usually are sufficient for plain imaging and shows a characteristic pattern with involvement of the middle third of the clavicle, with anterior and superior tilting of the sternal half and a smaller acromial portion, lack of reactive callus, and bulbous appearance of the lateral segment at the pseudarthrosis without tapering of the bone ends. The CTscans and particularly the 3-D (three dimensional) reconstruction images are being widely used recently as accurate, noninvasive diagnostic tools and play a vital role in reaching a diagnosis and formulating a treatment plan. MRI may be indicated in occasional cases (like thoracic outlet syndrome) to determine the extent of the fibrous union, the location of the great vessels, and the space available within the thoracic outlet.

Treatment of this condition varies. When patients are asymptomatic and have no functional disability, some authors do not recommend any treatment<sup>12-13</sup>. Conversely it has also been reported in literature that, if treated conservatively, the deformity can increase and lead to instability of the shoulder girdle

and to neuro-vascular complications. Surgical treatment is recommended for symptomatic patients with dysfunction of the arm, when patients are embarrassed by the cosmetic appearance of the lump and to prevent thoracic outlet syndrome in the future. The optimal age for surgery is controversial. Many workers recommend that surgery be done between 3 and 4 years of age. Operative intervention involves resection of the pseudarthrosis, with or without bone graft interposition, and internal<sup>11,14</sup> or external fixation<sup>2</sup>. Histopathological analysis of resected specimen confirms the diagnosis of a true congenital pseudarthrosis of the clavicle by revealing features of false joint with the ends of the clavicle covered with hyaline cartilage caps. The appearance of the cartilaginous caps is considered equivalent to that of developing physes and were proven to be adding new bone to the bony ends<sup>15</sup>. These observations are also supporting the hypothesis that the pseudarthrosis is caused by failure of two ossification centers to fuse, as has proposed by many workers in literature<sup>7</sup>.

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