

Carpal Coalitions and Metacarpal Synostoses: A Review

HAND
2016, Vol. 11(3) 271–277
© American Association for
Hand Surgery 2016
DOI: 10.1177/1558944715614860
hand.sagepub.com

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Abstract

Background: Carpal coalition and metacarpal synostosis are uncommon congenital anomalies of the carpus and hand. **Methods:** A comprehensive review of the literature was performed to help guide surgical and non-surgical treatment of carpal coalition and metacarpal synostosis. **Results:** The embryology, epidemiology, medical and surgical management, and associated outcomes are detailed. **Conclusions:** Most patients with these disorders will likely benefit from conservative measures. Surgery should be considered in patients with pain and limitations in wrist and hand function.

Keywords: carpal coalition, synostosis, hand coalitions, congenital, lunotriquetral coalition

Introduction

Congenital carpal coalitions are often found incidentally after radiographic evaluation following trauma. As such, the true incidence of carpal coalitions is unknown, with several case series and reports providing the most current available data. In contrast to isolated congenital carpal coalitions, those occurring in association with syndromes have been well defined and demonstrate a wide array of anatomic variability. The coalition between the lunate and triquetrum has been the best described in the literature secondary to its higher incidence than other coalitions. As most isolated congenital coalitions are asymptomatic, treatment is often not required.

Although congenital carpal coalition is well described as a failure of segmentation, metacarpal synostosis is best described as a failure of differentiation causing a union between 2 adjacent metacarpals. Metacarpal synostosis has been most described between the ring and small fingers and oftentimes is symptomatic.

In an effort to help guide identification and treatment of these rare entities, this article will review relevant classification systems, associated syndromes, and treatment options that have been described in the literature.

Embryology

Embryologically, carpal coalitions arise from a failure of separation of the cartilaginous interzone of adjacent carpal bones. The interzone is made up of 3 layers: 2 denser zones that form the articular cartilage of the 2 bones and a central loose layer that becomes the synovium and intracapsular

structures.^{36,48,53} The extent of coalition between carpal bones is determined by the amount of lack of development of the central zone. Patients with no development of the central zone will have a complete coalition whereas those with incomplete development will have a partial coalition. When describing a partial or incomplete bony union between adjacent carpal bones, many authors prefer the term *incomplete coalition*.^{19,36,52} This theory applies specifically to carpal bones in the same row and is not supported in coalitions of the pisiform and hamate. This joint is hypothesized to coalesce due to ossification of the distal portion of the flexor carpi ulnaris or through metaplasia of the pisohamate ligament into bone resulting in synostotic fusion and not true coalition.^{5,7,8,35,43,49}

Unlike carpal coalitions, isolated metacarpal synostosis is thought to be due to a sporadic inheritance pattern causing failure of differentiation between adjacent metacarpals.^{4,32,56} However, recent data suggest that genetic inheritance may play a larger role than previously thought. With the help from genetic sequencing and splicing, an X-linked recessive inheritance pattern has been described. This pattern involves a nonsense mutation in exon 3 of FGF16 on chromosome Xq21.1.³²

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Table 1. Minnaar Classification.

Type I	Incomplete fusion resembling a pseudarthrosis
Type II	Fusion with a notch of varying depth
Type III	Complete fusion of lunate and triquetrum alone
Type IV	Complete fusion associated with other carpal anomalies

Epidemiology

As most patients with congenital carpal coalitions are asymptomatic and found incidentally, the true incidence of congenital carpal coalitions is unknown. The incidence of congenital carpal coalitions in the literature has varied from as low as 0.1% in Caucasians to as high as 8% in certain African tribes.^{7,20,24,48} The most frequent coalition described is between the lunate and triquetrum, and is twice as common in females than males.²⁰

Metacarpal synostosis is relatively rare with a wide variation in patient presentation.^{4,16,23,40} As previously mentioned, the fourth and fifth metacarpal is most often affected. The incidence of metacarpal synostosis has ranged in the literature from 0.02% to 0.07%. As many names have been attributed to what has now been described as metacarpal synostosis, this range may not be all-inclusive.⁴

Classification Systems

Several classification systems have been described in an effort to help guide treatment of these 2 rare congenital anomalies. Case reports describing carpal coalitions date back to as early as 1779, with several subsequent reports following.^{12,55}

DeVilliers Minnaar defined the first carpal coalition classification system in 1952. The Minnaar classification was developed when describing 12 cases of congenital coalition of the lunate and triquetrum of the South African Bantu.¹² The classification includes 4 subtypes as described by Minnaar, and has been extended to help describe other carpal coalitions (see Table 1). Although this scheme has been used extensively for both luno-triquetral coalitions, as well as many others, it has several shortcomings. These limitations mainly apply to the identification and classification of associated anomalies.^{5,28} Specifically, anomalies are often seen with Minnaar types I and II and are not necessarily restricted just to the hand, as they can be seen in the lower extremity.^{6,28} Furthermore, it does not address the significant variability seen in nonosseous coalitions. To remedy these pitfalls, a classification was proposed by Burnett et al, who describe coalitions as either nonosseous or osseous.⁵ As incomplete coalitions are often more symptomatic, this classification likely has more clinical significance than the aforementioned system.^{5,45,48,49,50,58}

Despite several classifications being described in the literature, none have truly captured the involvement or lack of

involvement of associated syndromes. Isolated coalitions without syndrome involvement have been described among the entire carpus and most often only involve 2 bones within the same row (see Figures 1a and 1b).^{7,20,29,33,36,41,45,47,49-51,58-60} In contrast, syndromic involvement often includes multiple bones in various patterns (see Figure 1c).

Despite the rare incidence of metacarpal synostosis, several classification systems have been described.⁴ Buck-Gramcko and Wood described a classification scheme in 1993 (see Table 2).⁴ The classification uses the length of synostosis to characterize these rare entities in to 3 subsets. This scheme is simple to use; however, it does not offer guidelines for treatment. Thus, Foucher et al developed a different classification scheme in 2001.¹⁶ This newer classification uses the shape of the synostosis, degree of metacarpal hypoplasia, deformity of the digits distal to the synostosis, web involvement, and growth direction of the epiphysis.

Isolated Carpal Coalitions

An isolated complete coalition between the lunate and triquetrum remains the most common coalition described in the literature.^{10,50} As these coalitions tend to be asymptomatic, they are often discovered incidentally and do not require any treatment. In contrast, incomplete coalitions have recently been proven to be more symptomatic than their complete counterpart.^{48,52,49,50,58} This has been attributed to the development of a thin layer of articular cartilage, which is prone to degenerative wear and sometimes even fracture.⁵⁰ In patients with symptomatic incomplete coalitions, lunate and triquetrum fusion provides acceptable results with little wrist dysfunction.^{52,24,50,58}

Most isolated coalitions occur within the same carpal row. However, case reports and series have described coalitions between the hamate and pisiform.⁸ Although the first case reported was asymptomatic, most subsequent reports have portrayed ulnar-sided symptoms including pain and ulnar nerve paresthesias.^{2,5,18,35,49} Like incomplete coalitions, the nonosseous coalition that can develop between the hamate and pisiform has been reported to be more symptomatic than its osseous counterpart, especially in regard to ulnar neuropathy.⁵ Similarly, nonosseous hamate and pisiform coalitions are predisposed to degenerative arthritis and fracture.^{6,18} Like with many coalitions, nonoperative measures such as immobilization should be tried first. Should conservative measures fail, excision of the pisiform and the underlying coalition should be entertained.^{5,35,49}

Metacarpal Synostosis

Isolated metacarpal synostosis can also be partial or complete. Typically, the synostosis is located between the ring and little finger metacarpals, but has also been described between the middle and ring fingers (see Figures 2a and

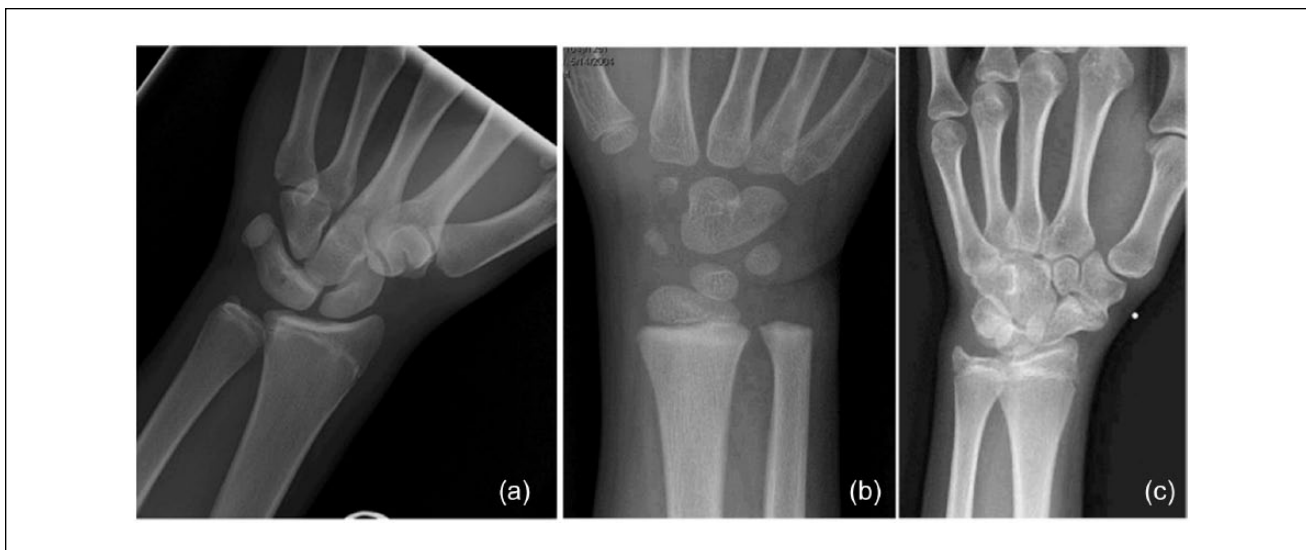


Figure 1. (a) PA radiograph hand depicting a lunate-triquetrum coalition with a very subtle cleft between the 2 bones distally. The trapezium and trapezoid may have a partial coalition as well. (b) PA radiograph demonstrates a capitata-hamate coalition. (c) PA radiograph with multiple carpal coalitions, including capitata-hamate and scaphoid-trapezium. Note. PA = posteroanterior.

Table 2. Buck-Gramcko and Wood⁴ Classification.

Type I	Coalition only at the base of the metacarpal
Type II	Synostosis extends up to half the length of the metacarpal
Type III	Synostosis extends more than half the length of the metacarpal

2b).^{4,23,27} This condition can be disabling because the affected finger is held in abduction. The abduction deformity limits finger motion and can cause the digit to catch on pockets. Should the deformity occur between the middle and ring, grasping objects becomes difficult, as they often will fall through the abduction deformity.^{4,23,27} To quantify the abduction deformity, the senior author attempted to describe the extent of deformity using standard posteroanterior (PA) radiographs of the hand with the affected finger in maximum adduction.²³ When the deformity is between the ring and small fingers, the middle finger is used as a reference point. If the middle and ring fingers are affected, the angle between the proximal phalanges is used.²³

Because of the variability in metacarpal synostoses, treatment can vary substantially. However, most surgical techniques involve splitting the synostosis and then filling the newly created space with a spacer to separate the metacarpals.^{4,16,23,27,40} The senior author's preferred filler is coralline hydroxyapatite (Interpore, Biomet, Parsippany, New Jersey).²³ It is important to place the graft proximal to the growth plates to avoid growth arrest. This technique corrects the abduction deformity but does not take in to account the hypoplastic deformity of the fingers.²³ As such, some

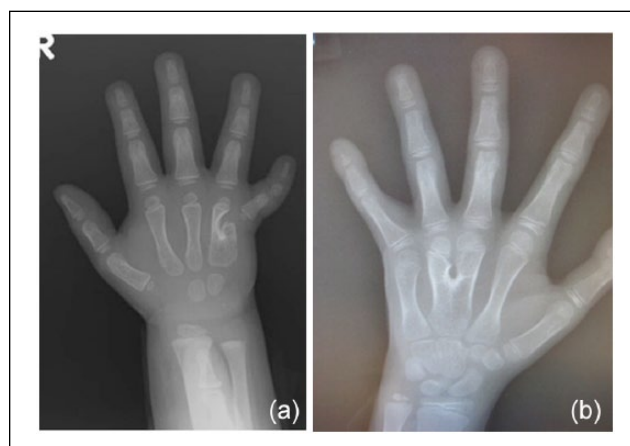


Figure 2. (a) PA radiograph of the hand with a metacarpal synostosis between the ring and small metacarpals. Note the abduction of the small finger. (b) PA radiograph with the metacarpal synostosis between the middle and ring finger metacarpals. Note. PA = posteroanterior.

impairment in motion and function will remain. Surgery should be considered on a case-by-case basis, as not all synostoses are amenable to separation with spacer placement.

Associated Syndromes

Several syndromes have been identified that are associated with carpal coalitions. These syndromes include arthrogyria, diastrophic dwarfism, dyschondrosteosis, Ellis-van

Creveld syndrome, fetal alcohol syndrome, hand-foot-genital syndrome, Holt-Oram syndrome, Nievergelt's syndrome, oto-palato-digital (OPD) syndrome, symphalangism, and Turner syndrome. Syndromic coalitions often cross carpal rows, involve multiple carpal bones, and are seen with other anatomic anomalies both within the appendicular skeleton and other organ systems.^{10,47,52} Although syndromic coalitions are more involved, few, if any, require surgical intervention.

Arthrogryposis Multiplex Congenita

Arthrogryposis is a constellation of clinical findings with the most notable being multiple congenital contractures of differing body areas.^{1,34} The incidence of arthrogryposis is approximately 1 in every 3000 live births and is thought to be due to a limitation of motion in utero.³ The exact mechanism for this is unknown although some have attributed it to antibodies inhibiting fetal acetylcholine receptors.

Patients with arthrogryposis and upper extremity abnormalities will present with a rigid deformity of the wrist and hand. The deformity is often a flexed and ulnarly deviated hand, which routinely fails to respond well to conservative measures.¹⁴

Carpal coalitions occur frequently in arthrogryposis and have substantial variability. The proximal row is more commonly involved early and then is often followed by intercarpal involvement. Authors have theorized this pattern of involvement as being acquired instead of congenital. Given the extensive nature of the coalitions, the midcarpal joint is often eliminated.¹⁴ This can often be attributed to a coalition between the scaphoid and capitate. A coalition of these two carpal bones complicates the traditional surgical treatment of a proximal row carpectomy. Instead, previous authors have recommended a biplanar osteotomy to correct the flexion and ulnar deviation of the carpus prior to the toddler age reaching 5 years of age.¹⁴

Diastrophic Dwarfism

Diastrophic dwarfism is an autosomal recessive form of short-limbed dwarfism. The common mutation involves the sulfate transporter protein found on chromosome 5 and is more common in patients of Finnish descent. Similar to arthrogryposis, it has been hypothesized that carpal coalitions associated with this syndrome may be acquired instead of truly congenital. This is in part due to the higher incidence of coalitions, as patients get older. The radiographic appearances of carpal bones appear deformed and more mature than their normal counterparts.⁴⁷ Management depends on the symptoms and expectations of the patient and their parents.

Dyschondrosteosis

Dyschondrosteosis is a rare inherited mesomelic dwarfism associated with bilateral Madelung deformities. Patients may also have associated variable carpal coalitions with limited forearm and wrist motion. Treatment options are often tailored toward the wrist deformity if symptomatic.

Ellis-van Creveld

Ellis-van Creveld syndrome is a rare autosomal recessive disorder with genetic defects in the EVC1 and EVC2 gene on chromosome 4p16.^{42,46} The syndrome is often referred to as chondro-ectodermal dysplasia. Characteristic features include short limb dwarfism and in over half of cases congenital heart defects. Other defects often include skin, teeth, hair, and nails.^{13,17,42,46,47} The incidence of the syndrome is 1 in 60 000 live births and affects boys and girls equally. Hand and wrist findings may include but are not limited to capito-hamate coalition, postaxial polydactyly, metacarpal synostosis, and clinodactyly.¹³ Although the patient may have significant carpus and hand involvement, treatment is often not necessary for the carpal coalition.

Fetal Alcohol Syndrome

Fetal alcohol syndrome has been recognized in children of mothers using alcohol during their pregnancy. Several consistent malformations have been described including those affecting the upper extremity. These can include radio-ulnar synostoses and capito-hamate coalitions either unilateral or bilaterally.³¹ Treatment is often conservative and based on symptoms.

Hand-Foot-Genital Syndrome

Hand-foot-genital syndrome is an autosomal dominant inherited syndrome characterized by hand and foot deformities with associated urogenital defects.⁵⁴ Hand and wrist manifestations include hypoplastic hypothenar eminence, limited thumb opposition attributed to limited flexion, and carpal coalitions. Coalitions have been described between the scaphoid and trapezium.^{22,25,30,54}

Holt-Oram Syndrome

Holt-Oram syndrome is an autosomal dominant inherited syndrome that affects the upper extremities and the heart. Patients with Holt-Oram syndrome will likely have an absent radius, atrial septal defect, and an abnormally shaped scaphoid with associated carpal coalitions.⁴⁷ Some authors have reported a narrowing of the scaphoid and trapezium joint space with accessory carpal bones.⁴⁷

Nievergelt's Syndrome

Nievergelt's syndrome is a rare form of mesomelic dwarfism. Although it often has an autosomal dominant inheritance, sporadic inheritance has also been documented. Classic features include limited motion of the elbow and forearm with associated carpal coalitions⁴⁴; Similar manifestations may also be seen in the feet including clubfeet and metatarsal synostosis.⁴⁴

OPD syndrome

OPD syndrome is a constellation of abnormalities affecting the ears, palate, and skeleton. The defining skeletal dysplasia affects the hands and feet predominantly. Carpal bones may demonstrate a deformed appearance.^{38,47} Although not specific for OPD, authors have described the trapezoid taking a comma-like shape with an associated transverse-oriented capitate. The scaphoid may also be involved but as with other syndromes, carpal coalitions may be highly variable.^{38,47}

Symphalangism

Symphalangism is an autosomal dominant inherited disorder described as having interphalangeal joint fusions with associated carpal coalitions.^{9,21,39,57} Symphalangism is often referred to as either proximal or distal, which refers to the distal or proximal interphalangeal joint. The proximal form is more common. Three forms have been described in the literature: Type 1 refers to true symphalangism without additional skeletal abnormalities, type 2 refers to symphalangism associated with symbrachydactyly, and type 3 refers to symphalangism with syndactyly.¹⁵ Fingers are often affected from an ulnar to radial pattern with the small finger being the most commonly affected.^{11,15,39}

Other additional upper extremity deformities have been associated with symphalangism and include brachydactyly, camptodactyly, clinodactyly, syndactyly, and radiohumeral fusions. Several carpal coalitions have also been described and include the triquetrum-hamate (most common), capitate-hamate, capitate-trapezium, triquetrum-lunate, and scaphoid-trapezium.^{39,57}

Patients often will present with the inability to move their proximal or distal interphalangeal joints with no visible joint creases. Radiographs will often demonstrate the absence of interphalangeal joints (skeletal immature patients may have a pseudo-joint with a cartilaginous bridge that has yet to ossify). Although patients lack interphalangeal joints, hand function is often unimpaired requiring little to no treatment.^{21,26,47}

Turner Syndrome

Turner syndrome is a chromosomal disorder marked by a single X chromosome. The disorder is manifested across

multiple systems but also typically affects the carpus. Previously, authors have described the typical anomaly seen in Turner syndrome as having deformed proximal carpal bones with a coarse trabecular pattern.³⁷ Others have described associated Madelung's deformity with a lunate-triquetrum coalition, but this is not specific to Turner syndrome.

Conclusion

Carpal coalitions and metacarpal synostosis represent rare congenital anomalies seen in both isolation and with associated syndromes. Both disorders represent a spectrum ranging from complete to incomplete coalitions/synostosis. Although most patients with these disorders will likely benefit from conservative measures, surgery should be considered in patients presenting with pain and limitations in wrist and hand function.

Ethical Approval

Permission has been granted to use photos and radiographs of specific patients.

Statement of Human and Animal Rights

This article does not contain any studies with human or animal subjects.

Statement of Informed Consent

All personal and identifying information for radiographs presented in this article have been removed.

Declaration of Conflicting Interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

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