Bilateral scaphotrapezium coalition with bilateral scaphoid nonunion in a patient with Klippel–Feil syndrome: a case report

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Introduction

The purpose of this case report is to discuss the evaluation and management of scaphoid nonunion in patients with congenital musculoskeletal syndromes. Klippel–Feil syndrome (KFS) is a rare disorder characterized by the presence of congenital synostosis of some or all cervical vertebrae [26] caused by a failure in the normal segmentation of the cervical vertebrae during the third to the eighth week of gestation [25]. The classic clinical triad of KFS consists of a short neck, low posterior hairline, and limited neck range of motion due to cervical synostosis [26]. Abnormalities of the hand have been associated with KFS [2, 4, 8, 15, 25] and are predominantly preaxial [2, 4, 15, 25]. One instance of a postaxial anomaly in a patient with KFS has been reported [2]. A prior identification of capitohamate coalition in a patient with Klippel–Feil syndrome was suggested to be a manifestation of a new clinical syndrome [14]. Additional carpal coalitions have not been reported nor have scaphoid abnormalities secondary to the carpal coalitions been described that are associated with Klippel–Feil syndrome.

Case Report

A 14-year-old left-hand dominant male with Klippel–Feil syndrome, manifested by cervical spine neck webbing and bilateral mildly hypoplastic thumbs, presented with 1 year of left wrist pain following a minor fall while playing soccer (Fig. 1) [15, 26]. He had experienced wrist pain during activities of daily living and was treated with splinting. He was referred to a local hand surgeon and evaluated with radiographs and an MRI due to continued pain and loss of range of motion. He was diagnosed with a scaphoid nonunion of the left wrist. Despite conservative treatment, he continued to experience mild pain with lifting, turning door knobs, and playing volleyball. The right wrist was asymptomatic and had no history of injury.

On examination, the radial pulse was absent bilaterally to Doppler signal. He experienced significant tenderness at the base of bilateral thumbs; a scaphoid shift test was painful. Wrist range of motion of the affected wrist was limited (extension/flexion, 30/70° (70/70° contralateral); radial/ulnar deviation, 10/20° (30/35° contralateral); palmar/radial abduction 50/50° bilaterally) as was his grip strength of 16 kg (24 kg contralateral). He maintained full range of motion of his digits with normal thumb opposition. He had thumb hypoplasia with narrowing of his thumb–index web space, hypoplastic intrinsic thenar muscles, and a mild degree of metacarpophalangeal joint laxity [10]. Hand and wrist radiographs were obtained along with a CT of the left wrist revealing scaphotrapezium coalition, scaphoid nonunion, and dorsal intercalated segment instability (DISI) deformity (Figs. 2 and 3). Due to the unusual radiographic findings of the left wrist and the presence of other congenital abnormalities, the right wrist was evaluated with radiographs. The right wrist demonstrated similar abnormalities, despite being asymptomatic (Fig. 2).

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Fig. 1 a, b General appearance of the 14-year-old male consistent with Klippel–Feil syndrome. The classic clinical triad of KFS consists of a short neck, low posterior hairline, and limited neck range of motion

Treatment options for scaphoid nonunion are well established [11, 12, 19, 23, 24, 27, 28]. Intraoperatively, an established scaphoid nonunion of the left wrist was identified with no soft tissue attachment to the proximal pole fragment (Fig. 4). The patient was treated with successful scaphoidectomy and four-corner arthrodesis. A four-corner fusion was chosen to prevent potential radio-carpal abutment with radial deviation because of the radial-sided coalition, but a proximal row carpectomy could have also been performed.

Twenty-four months following operation, the patient was experiencing no pain unless his wrist was overstressed. He experienced no limitations in his daily or recreational activities. Wrist range of motion was improved (extension/flexion, 46/25° (67/65° contralateral), radial/ulnar deviation, 18/18° (25/48° contralateral)) as was grip strength of 37 kg (48 kg contralateral). Postoperative radiographs revealed solid bony fusion along with normal postoperative findings (Fig. 5). His right wrist remained asymptomatic, and it has been reevaluated yearly with radiographs and clinical examinations.

Discussion

Carpal coalitions have been described between nearly all of the carpal bones with lunotriquetrum followed by capitohamate being the most frequent [9, 21]. Carpal coalitions associated with syndromes tend to involve more than two carpals, frequently involve the scaphoid, and have fusions across the carpal rows [3, 21]. More complex nonsyndromic congenital carpal coalitions such as scaphotrapeziotrapezoid fusion and congenital fusion of the scaphoid, trapezium, trapezoid, and capitate have also been observed [1, 29] in the general population. Carpal coalitions may place the wrist at greater risk of scaphoid fracture and nonunion [10].

Approximately 5–10% of the 345,000 scaphoid fractures annually in the USA result in nonunion [18, 20]. Although the overall prevalence of scaphoid nonunion is low [22], the risk of osteoarthritis in patients with nontreated scaphoid nonunion nears 100% [16, 18]. The presented case suggests

Fig. 2 a–d Posterior–anterior and lateral radiographs of the left symptomatic (a, b) and right symptomatic (c, d) wrists demonstrating a scaphoid nonunion with a dorsal intercalated segmental instability pattern and coalition of the scaphotrapezium joint

Fig. 3 **a, b** Sagittal and coronal CT reconstructions of the left wrist demonstrate a scaphotrapezium coalition and nonunion of the scaphoid

that underlying congenital malformations of the wrist may place patients at higher risk for nonunion. In patients with musculoskeletal syndromes, radiographs of the contralateral wrist and CT scan of the symptomatic wrist provide additional insight into the patient's presentation. In the above case, radiographs revealed either bilateral scaphoid nonunion or congenital bipartite scaphoid of the asymptomatic wrist with DISI deformity suggesting potential baseline instability. As a result, the patient's symptomatic side was treated aggressively to reduce wrist instability and avoid the development of osteoarthritis.

The findings of bilateral scaphoid nonunions and bilateral carpal coalitions involving proximal and distal rows with bilateral thumb hypoplasia are very unusual, although radial-sided carpal abnormalities can be associated with thumb hypoplasia. The presence of a contralateral asymptomatic scaphoid nonunion in the absence of a history of trauma leads one to consider the possibility of a congenital bilateral bipartite scaphoid. There are several case reports of bipartite scaphoid, including bilateral bipartite scaphoids suggestive of a congenital etiology [7, 13, 17]. It remains possible that this patient sustained trauma to both wrists and developed bilateral scaphoid nonunion. However, the patient denies any trauma to the asymptomatic side and had only a minor trauma to the affected side. Additionally, the intraoperative finding of having a completely free floating proximal scaphoid with no soft tissue attachments in a pediatric age patient is not consistent with most pediatric scaphoid nonunions [5, 6].

In conclusion, we report an unusual case of bilateral scaphoid nonunions, thumb hypoplasia, and carpal coalitions in a boy with KFS. In patients with musculoskeletal syndromes who present with wrist pain, it is important to carefully examine both wrists and evaluate the radiographic studies to determine if carpal anomalies may be contributing to their wrist pain. This case reveals the value in obtaining films of the contralateral wrist in an effort to diagnose baseline instability of the wrist. The case supports the position that carpal coalition may place scaphoid fractures at higher risk of nonunion. Treatment of the

carpal conditions in these patients requires individualization and a systematic and detailed approach that is thoroughly discussed preoperatively. It is also important to recognize that the wrist abnormalities may be secondary to long-

Fig. 4 **a, b** Intraoperative volar (**a**) and dorsal (**b**) approaches to the scaphoid nonunion. Dorsal approach reveals a partially resected distal pole and a four-corner arthrodesis using iliac crest bone graft and fixed with a mini-Spider plate (KMI, San Diego, CA, USA)

Fig. 5 **a, b** Postoperative posterior–anterior and lateral radiographs revealing solid fusion of four-corner arthrodesis

standing congenital abnormalities prompting symptomatic treatment.

Conflicts of interest The authors declare that they have no conflict of interest.

References

- Carlson DH. Coalition of the carpal bones. *Skeletal Radiol.* 1981;7(2):125–7.
- Chemke J, Nisani R, Fischel RE. Absent ulna in the Klippel–Feil syndrome: an unusual associated malformation. *Clin Genet.* 1980;17(2):167–70.
- Cockshott WP. Carpal fusions. *Am J Roentgenol Radium Ther Nucl Med.* 1963;89:1260–71.
- da Silva EO. Preaxial polydactyly and other defects associated with Klippel–Feil anomaly. *Hum Hered.* 1993;43(6):371–4.
- Elhassan BT, Shin AY. Scaphoid fracture in children. *Hand Clin.* 2006;22(1):31–41.
- Elhassan BT, Shin AY, Kozin SH. Scaphoid fractures in children. In: Shin AY, editor. *Scaphoid fractures and nonunions*. Monograph series 37. Rosemont: American Academy of Orthopaedic Surgeons; 2007.
- Faulkner DM. Bipartite carpal scaphoid. *J Bone Jt Surg.* 1928;10:284–9.
- Greenspan A, Cohen J, Szabo RM. Klippel–Feil syndrome. An unusual association with Sprengel deformity, omovertebral bone, and other skeletal, hematologic, and respiratory disorders. A case report. *Bull Hosp Joint Dis Orthop Inst.* 1991;51(1):54–62.
- Ingram C, Hall RF, Gonzalez M. Congenital fusion of the scaphoid, trapezium, trapezoid and capitate. *J Hand Surg Br.* 1997;22(2):167–8.
- James MA et al. The association of radial deficiency with thumb hypoplasia. *J Bone Joint Surg Am.* 2004;86-A(10):2196–205.
- Jeon I et al. Clinical result of operative treatment for scaphoid non-union in the skeletally immature: percutaneous versus open procedure. *Hand Surg.* 2008;13(1):11–6.
- Kawamura K, Chung K. Treatment of scaphoid fractures and nonunions. *J Hand Surg Am.* 2008;33(6):988–97.
- Kim SB, Kim WS, Chung WY, Jeon TS, Bae IT. Bilateral bipartite carpal scaphoid: a case report. *J Korean Orthop Assoc.* 2005;40(5):614–6.
- Kozlowski K, Sillence D, Taylor F. Short stature, mental retardation, craniosynostosis, Klippel–Feil syndrome, Scheuerman kyphosis, rib gaps and other distinctive skeletal and genital anomalies. A new syndrome? *Pediatr Radiol.* 1993;23(6):442–5.
- Lee WT et al. Thenar hypoplasia in Klippel–Feil syndrome due to aberrant radial artery. *Pediatr Neurol.* 1995;13(4):343–5.
- Lindström G, Nyström A. Incidence of post-traumatic arthrosis after primary healing of scaphoid fractures: a clinical and radiological study. *J Hand Surg Br.* 1990;15(1):11–3.
- Louis DS et al. Congenital bipartite scaphoid—fact or fiction? *J Bone Joint Surg Am.* 1976;58(8):1108–12.
- Mack G et al. The natural history of scaphoid non-union. *J Bone Joint Surg Am.* 1984;66(4):504–9.
- Merrell G, Wolfe S, Slade JF. Treatment of scaphoid nonunions: quantitative meta-analysis of the literature. *J Hand Surg Am.* 2002;27(4):685–91.
- Osterman A. Scaphoid non-union. *Hand Clin N Am.* 1988;4:437–55.
- Poznanski AK, Holt JF. The carpals in congenital malformation syndromes. *Am J Roentgenol Radium Ther Nucl Med.* 1971;112(3):443–59.
- Sehat K, Bannister G. The prevalence of established scaphoid fracture non-union found on incidental radiography. *Injury.* 2000;31(4):275–6.
- Steinmann S, Adams J. Scaphoid fractures and nonunions: diagnosis and treatment. *J Orthop Sci.* 2006;11(4):424–31.
- Steinmann S, Bishop A. A vascularized bone graft for repair of scaphoid nonunion. *Hand Clin.* 2001;17(4):647–53. ix.
- Thomsen M et al. Congenital limb deficiencies associated with Klippel–Feil syndrome: a survey of 57 subjects. *Acta Orthop Scand.* 2000;71(5):461–4.
- Tracy MR, Dormans JP, Kusumi K. Klippel–Feil syndrome: clinical features and current understanding of etiology. *Clin Orthop Relat Res.* 2004;424:183–90.
- Tu Y et al. Treatment for scaphoid fracture and nonunion—the application of 3.0 mm cannulated screws and pedicle vascularised bone grafts. *Injury.* 2008;39 Suppl 4:96–106.
- Waitayawinyu T et al. Scaphoid nonunion. *J Am Acad Orthop Surg.* 2007;15(5):308–20.
- Weinzweig J et al. Congenital synchondrosis of the scaphotrapezio-trapezoidal joint. *J Hand Surg Am.* 1997;22(1):74–7.