





# Congenital aplasia of the scaphoid bone

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Congenital aplasia of the scaphoid bone without thumb or radial hypoplasia is a rare condition. Scaphoid hypoplasia and aplasia has been defined with syndromes such as radial hemimelia, absence of thumb, vertebral defects, anal atresia, cardiac anomalies, tracheoesophageal fistula, renal anomalies, and limb abnormalities (VACTERL), Holt-Oram syndrome (heart defects and upper extremity anomalies), and thrombocytopenia-radius anomalies (TAR). Scaphoid anomalies are often seen associated with absence of thumb or radius. In the literature, there are seven case reports with isolated scaphoid aplasia without other congenital abnormalities, but none of them are current.<sup>[1-7]</sup> Herein, we present a very rare case diagnosed in the pediatric age group with hypoplasia of the biceps brachii ipsilateral to scaphoid aplasia.

## CASE REPORT

A 12-year-old male patient was applied to the orthopedics and traumatology outpatient clinic

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## ABSTRACT

Congenital aplasia of the scaphoid bone without thumb or radial hypoplasia is a rare condition. In the literature, there are seven case reports of congenital scaphoid aplasia without other congenital abnormalities, but none of them are current. Scaphoid hypoplasia and aplasia have been defined with syndromes such as radial hemimelia, absence of thumb, vertebral defects, anal atresia, cardiac anomalies, tracheoesophageal fistula, renal anomalies, and limb abnormalities (VACTERL), Holt-Oram syndrome (heart defects and upper extremity anomalies), and thrombocytopenia-radius anomalies (TAR). This is a very rare case diagnosed in the pediatric age group with hypoplasia of the biceps brachii ipsilateral to scaphoid aplasia. Herein, a 12-year-old boy with unilateral scaphoid agenesis is presented, and its clinical and imaging findings as well as the treatment are discussed.

**Keywords:** Aplasia, congenital, scaphoid.

12 months ago with right wrist weakness and limited range of movement. After the physical examination and radiographs of the patient, he was diagnosed with scaphoid agenesis. There were no features in the history, no additional systemic disease, no syndromic facial appearance, and the patient's intelligence was normal.

The right wrist differed from the left side in terms of thenar atrophy, palmar creases, and distal wrist crease (Figure 1). On the right side, hypoplasia of the biceps brachii was seen (Figure 2). There was no tenderness and pain in the first carpometacarpal and radiocarpal joints on palpation. Right radial pulse was non-palpable, right ulnar pulse was palpable, and there was a delay on the right in Allen's test. Wrist instability tests were negative. There was no neurological deficit. Bilateral upper extremities were of equal length. Thumb and other finger joint ranges of motion were normal and right wrist movements were diminished, particularly in dorsiflexion. The joint range of motion measurement



**Figure 1.** Morphological images of the hands.



**Figure 2.** Hypoplasia of the biceps brachii.

values and grip strength of the patient are presented in Table 1.

In the radiographic examination, there was no scaphoid bone on the right side, the alignment and morphology of the other carpal bones were normal (Figure 3). The carpal bones on the left had a normal appearance. The trapezium had a dysplastic appearance on the right, lost its pyramidal feature, and its height was measured as 10.6 mm, on the left, it was measured as 14.8 mm. The right radius styloid had a hypoplastic appearance, and the distal radioulnar joint was normal. Carpal height ratio was measured as 0.36 on the right wrist and 0.59 on the left, 38% less on the side without scaphoid. There were no degenerative changes in the intercarpal and radiocarpal joints. Magnetic resonance imaging (MRI) confirmed the absence of scaphoid bone in the right wrist. Other carpal bones were in normal configuration. The morphology and signals of other carpal bones forming the wrist were normal. No pathology was detected in the periarticular tissues. Computed tomography (CT) showed that the scaphoid bone socket was smaller than expected and there was a millimetric ossific bone fragment on the volar side. It was thought that the ratio of the radius distal articular surface to the ulna articular surface may

have decreased. No significant deterioration was observed in the axis (Figure 4). On electromyography, partial dysfunction of the motor and sensory branches of the median nerve was reported, compatible with neuropathy. Brachial plexus MRI was performed and no pathology was detected. Surgical treatment was not planned for the patient with these findings. The patient is still under follow-up on a regular basis in the orthopedic outpatient setting.

The parents were informed that data from the case would be submitted for publication and gave their consent.

**TABLE 1**

Joint range of motion values and grip strength measurement		
	Right	Left
Range of motion		
Flexion	55	75
Extension	10	65
Ulnar deviation	10	30
Radial deviation	15	20
Grip strength (kg)		
Strength at Jamar	1.5	7.3
K-Pinch	10.6	25.2



Figure 3. X-ray images of both wrists.



Figure 4. Three-dimensional computed tomography images of the wrists.

## DISCUSSION

Scaphoid aplasia is rare. In the literature, there are seven cases with isolated scaphoid aplasia (Table 2).<sup>[1-7]</sup> The most recent publication on this subject is dated 2012, and our case was written to contribute to the current literature. In addition, our case is the youngest case in age among the cases published on this subject.

Davison<sup>[8]</sup> created a classification of six different types of scaphoid hypoplasia, ranging from mild hypoplasia to complex hypoplasia and aplasia affecting the forearm and fingers. He concluded that the defect occurred as a result of embryological damage affecting the skeleton in the proximal and distal sequence. However, he did not explain the possibility of the absence of the scaphoid associated with the normal thumb. In the Davison's classification,

the complete absence of the scaphoid has always been associated with severe hypoplasia and aplasia of the thumb. Our case is not similar to the Davison classification, as there was no associated longitudinal disorder.

In the classifications published after O'Rahilly,<sup>[9]</sup> the concept of intercalary deficiency has been largely abandoned; therefore, this classification that includes intercalary radial hemimelia is the only classification consisting of congenital malformation of our patient.<sup>[7,9]</sup>

Radial styloid hypoplasia of our patient was attributed to the absence of scaphoid contact, and thenar atrophy was attributed to median nerve entrapment neuropathy. In the literature, carpal tunnel syndrome (CTS) was seen in patients with isolated scaphoid agenesis; however, since all of these

**TABLE 2**  
Review of the literature about aplasia of the scaphoid bone

Author	Year	Age/Sex	Clinical features	Article title
Papanikolaou and Haddadin <sup>[2]</sup>	1972	22/M	Pain in the left wrist as a result of a fall on the outstretched hand.	Congenital absence of carpal scaphoid.
Srivastava and Kochhar <sup>[4]</sup>	1972	26/M	Pain in wrist at the limits of motion.	Congenital absence of the carpal scaphoid
Kuz and Smith <sup>[6]</sup>	1997	18/M	Pain after a history of a right wrist sprain	Congenital absence of the scaphoid without other congenital abnormality: a case report.
Patankar <sup>[3]</sup>	1998	28/M	Pain, swelling and stiffness of the left wrist following an injury	Bilateral congenital aplasia of the scaphoid
Chamay <sup>[1]</sup>	2001	66/M	Pain localized at the base of the thumb	Unilateral congenital aplasia of the scaphoid and dysplasia of the trapezium: a case report
Pancieria and Le Viet <sup>[7]</sup>	2008	45/M	Pain, swelling, and stiffness of both wrists without any history of injury or other joint complaints.	Intercarpal degenerative arthritis in adulthood as a late consequence of unilateral congenital aplasia of scaphoid: a case report
Gómez Fernández et al. <sup>[5]</sup>	2012	49/M	Mild pain in the wrist for about 20 years.	Unilateral congenital absence of the carpal scaphoid associated with dysplasia of the capitulum

patients were elderly patients, it was assumed that scaphoid agenesis did not cause CTS. As a defense, the absence of CTS was shown in a 15-year-old young patient reported by Radford.<sup>[10]</sup> However, our patient had CTS despite his young age. This raises the question of whether scaphoid agenesis could cause CTS. In addition, carpal collapse was observed in all cases reported in the literature, and our case is no exception.

In conclusion, our patient did not have pain in joint movements, but his right wrist movements were limited compared to the left side. There was no sign of carpal instability in the wrist. Although the congenital aplasia of scaphoid bone is not the risk factor for carpal instability, there is no study comparing congenital scaphoid aplasia and acquired scaphoid excision in terms of instability.

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