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Abductor digiti minimi opponensplasty for a patient with Cavanagh's syndrome: A case report

Ali Parsa (MD)[1], Farzad Omidi Kashani (MD)[1], Mohammad Hallaj Moghaddam (MD)[1], Abdolreza Malek (MD)[2], Ali Birjandinejad (MD)[1]

[1]*Orthopedic Research Center, Department of Orthopedic Surgery, Mashhad University of Medical sciences, Mashhad, Iran.*
[2]*Department of Pediatrics, Mashhad University of Medical Sciences, Mashhad, Iran.*

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ABSTRACT

Thumb hypoplasia only in the intrinsic thenar muscles is a rare condition; this defect might be accompanied with cardiac diseases (Holt-Oram syndrome), ocular anomalies, and vascular anomalies of the hand and wrist (Okihiro syndrome). In addition, this condition may be detected in hypereosinophilic syndrome (HES), which gives rise to other hand anomalies, as well. Unilateral or bilateral absence of isolated thenar muscles is another rare abnormality.
In this study, we presented a case of Cavanagh's syndrome in an eight-year-old boy with right-sided thenar hypoplasia who had difficulty in thumb opposition.
For the treatment, the patient underwent abductor digiti minimi (ADM) opponensplasty. After three years of follow-up, he regained thumb opposition and was symptom-free. Care must be taken to avoid misdiagnosis of thenar atrophy/hypoplasia with carpal tunnel syndrome in case of children.

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Introduction

Thumb plays a pivotal role in opposition and prehension movements; it is considered the most important part of the upper extremities. Intrinsic muscles of the thenar eminence are the key elements in complex movements of the thumb. Anomalies involving the thumb and thenar muscles could be debilitating and have adverse effects on the activities of an individual (1).

Thumb hypoplasia is a relatively rare anomaly constituting a wide range of defects; for instance, one or more of the tendons and muscles, bones, or ectodermal tissues of the thumb may be absent in thumb hypoplasia. The defects caused by this anomaly vary from a short thumb to the complete absence of the thumb. Some reported cases of this anomaly were associated with the absence of intrinsic muscles in the thumb (2).

The incidence of thumb hypoplasia only in the intrinsic thenar muscles is a rare condition; this defect might be accompanied with cardiac diseases (Holt-Oram syndrome) (3), ocular anomalies, and vascular anomalies of the hand and wrist (Okihiro syndrome) (4,5). In addition, this condition may be detected in hypereosinophilic syndrome (HES), which gives rise to other hand anomalies, as well. Unilateral or bilateral absence of isolated thenar muscles is another rare abnormality. According to the literature, there are several cases of the coincidence of thenar hypoplasia and other hand and wrist anomalies (6,7).

According to a study conducted in 1979, many cases of thenar hypoplasia, also known as Cavana-

***Corresponding author:** Ali Parsa.*
Orthopedic Research Center, Department of Orthopedic Surgery, Mashhad University of Medical sciences, Mashhad,Iran.
***E-mail:** aliparsadr@yahoo.com*
***Tel:** +989155056358*

gh's syndrome, manifest certain degrees of thumb hypoplasia in radiological examinations (8). However, the actual cases of Cavanagh's syndrome have been rarely reported in the literature. In this syndrome, there is an isolated thenar hypoplasia without any extrinsic muscle or systemic anomalies. In this study, we presented a case of Cavanagh's syndrome in an eight-year-old boy with right-sided thenar hypoplasia. For the treatment, the patient underwent opponensplasty.

Case report

A right-handed, eight-year-old boy presented to the department of orthopedic surgery at Ghaem Hospital, Mashhad University of Medical Sciences, in January 2014, with complaints of hand muscle wasting and weakness and difficulty in performing fine motor functions.

The patient had no history of trauma . On examination, partial atrophy of the right thenar muscle compared to the left thenar was clearly visible (Figures 1a and 1b). The thumbs were normal and symmetrical on both sides, and deep flexor of the thumb was palpable.

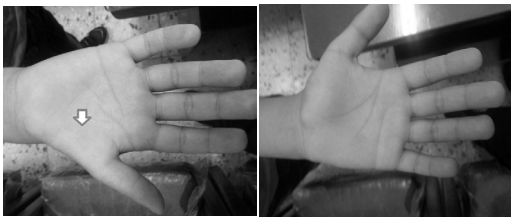

Figure 1a) Thenar hypoplasia (arrow) right hand. **b)** The normal side (left).

The thumbs' flexion and extension ranges of motion were symmetrical on both sides, but he was not able to oppose the thumb properly. For more accurate thumb apposition measurement, we evaluated the opposition ability by utilizing the Kapanji score. Kapanji score is a useful tool to evaluate the opposition and the counter-opposition of the thumb, this method does not require the measuring of angles, rather, the hand itself is used as the system of reference based on where on the hand the patient is able to touch with the tip of their thumb. The possible scores range from 1 to 10, score 1 indicating very limited ability to oppose the thumb (9). Kapanji score was 2 in the right hand and 8 in the left hand. Also, no sensory disorders were detected, and pulses were equal and strong on both sides. In radiological examination, there were no signs of hypoplasia or other malfunctions.

The patient was referred to a pediatric cardiologist, and medical examinations and echocardiography were performed. There were not any symptoms or signs of cardiovascular diseases.

Moreover, the patient was re-examined by a pediatrician, and there were no signs of any specific disorders.

In electro diagnostic testing, the patient was diagnosed with opponens pollicis dysfunction, and there were no additional problems.

For the treatment, the patient underwent abductor digiti minimi (Huber) opponensplasty. At first, a 7-cm medial incision was made at ulnar border of the hand, then abductor digiti minimi (ADM) was detached at the most distal part to obtain the maximum length origin of the ADM at pisiform; bone, as well as pisotriquetral and pisohamate ligaments remained intact. Pisiform bone in this procedure acts as the pivot point. The transferred ADM easily attached to the thumb metacarpophalangeal joint through a subcutaneous tunnel. Smooth movement without excessive tension was achieved (Figure 2). Rehabilitation therapy was performed after wound healing, and the function of opponens pollicis was normal after 18 months (Figure 3).

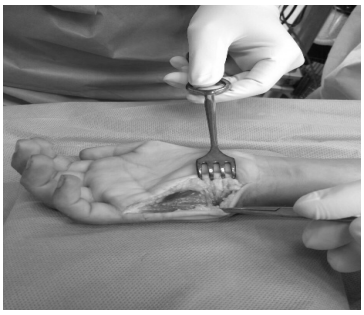

Figure 2. Abductor digiti minimi exposure.

Figure 3. Post-operative ability to oppose thumb.

Discussion

Several studies have confirmed that congenital abnormalities of the thumb could be extremely debilitating and cause severe functional problems. As previously mentioned, thumb hypoplasia is not a rare condition, and its association with other hand anomalies, such as thenar hypoplasia, has been confirmed in many cases. However, isolated thenar hypoplasia, also known as Cavanagh's syndrome, is a rare defect.

Several opposition transfers have been described in children including the abductor digiti minimi (ADM—Huber transfer) (10,11), exten-

sor pollicis longus (EPL) (12), palmaris longus (Camitz transfer) (11), and the flexor digitorum superficialis (FDS) (13). Of all these techniques, Huber transfer seems to be the most frequently used.

The first case of bilateral Cavanagh's syndrome was reported by Witt et al. in 1981. We found four other reports of Cavanagh's syndrome (1,6,4,15). In the present report and other similar studies, ADM opponensplasty was performed for the treatment of patients, while Tas et al. used the extensor indicis proprius transfer technique (15). We believe that the transfer of ADM is superior to flexor and extensor tendon transfers because the integrity of flexor and extensor mechanisms of fingers remains intact.

It is noteworthy that Cavanagh's syndrome might be confused with the carpal tunnel syndrome due to the inaccuracies in medical examinations. Thenar hypoplasia might be considered as thenar atrophy secondary to median nerve compression, especially if the focus is placed on nerve conduction velocity in the electrodiagnostic testing, without the consideration of electromyograms. Needle electromyography of median innervated muscles of thenar region (opponens pollicis and abductor pollicis brevis) can prevent this misdiagnosis.

On the other hand, idiopathic carpal tunnel syndrome rarely occurs in children, which should be considered in the differential diagnosis of Cavanaugh's syndrome. According to a study conducted by Potulska-Chromik et al., idiopathic carpal tunnel syndrome was detected in less than 10% of the cases, and 54% of the cases of carpal tunnel syndrome in children were secondary to congenital anomalies (16).

Conclusion

Our intent in presenting this case is to increase awareness of the congenital absence of thenar muscles and consider abductor digiti minimi transfer as a fine choice while caring for these patients.

Acknowledgement

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Conflict of Interest

The authors declare no conflict of interest.

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