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OLGU SUNUMU *Psikiyatri*

Restoration of function on bilateral absence of extensor pollicis longus and brevis: A case report

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SUMMARY

Thumb hypoplasia and radial dysplasia are two of the most common hand therapy requiring conditions among the congenital problems. However, any cases with congenital agenesis of bilateral thumb extensors, and their related rehabilitatitive therapy were not reported in the literature. The aim of this case report is to make the first contribution to the restoration of thumb extension function in a child with congenital absence of bilateral extensor pollicis longus (EPL) and extensor pollicis brevis (EPB) The case was a 5-year old girl and her family noticed that she had no thumb extension. Results of the tests performed to eliminate the other systemic problems were all in normal ranges. The patient underwent immobilization in a forearm cast with 20° wrist extension and full thumb extension and abduction for 6 weeks. During this period, early dynamic motion protocol was applied by a wire-rubber system splint. After removal of the splint, therapeutic ultrasound was applied and passive and active- assisted exercises gradually proceeded to resistive exercises through the 8th week. By the 8th week, occupational therapy with gradually increasing resistance was initiated while the patient was permitted to use both hands in daily activities after 4th month. Assessments on the post-operative 6th, 8th and 12th weeks revealed significant improvements in range of motion in the thumbs, grip and pinching strengths and hand functional level. Restoration of thumb extension with surgery and hand rehabilitation before school age provided optimal hand function and quality of life of this child.

Key words: *Thumb hypoplasia, hand function, hand therapy*

ÖZET

Bilateral ekstansör pollisis longus ve brevis yokluğunda el fonksiyonunun restorasyonu: Olgu raporu

Başparmak hipoplazisi ve radial displazi sıklıkla el terapisine yönlendirilen konjenital sorunlardandır. Ancak, literatüre bakıldığında başparmak ekstansör kaslarının bilateral konjenital yokluğuna ve bu durumun rehabilitasyonuna dair çalışmaların eksik olduğu göze çarpmaktadır. Bu olgu sunumunun amacı; bilateral konjenital ekstansör pollicis longus ve brevis yokluğu olan bir çocukta başparmak ekstansiyonunun restorasyonu ile ilgili literatüre katkıda bulunmaktadır. Beş yaşında bir kız çocuğu olan olgumuzun başparmak ekstansiyonu olmadığı ailesi tarafından fark edilmiştir. Bir ortopedist tarafından bilateral konjenital başparmak ekstansörlerinin yokluğu tespit edilmiş ve diğer sistemik sorunlar ekarte etmek üzere gerekli tetkikler yapılmıştır. Hastamız cerrahiden sonra 6 hafta boyunca 20° bilek ekstansiyonu ve tam başparmak ekstansiyon ve abduksiyonu sağlayan önkol splinti içerisinde immobilize edilmiştir. Erken dinamik hareket protokolüne göre 8. haftaya kadar pasif ve aktif-asistif egzersizler yapılmış, bu haftadan itibaren dirençli egzersizlere geçilmiştir. Yine 8. haftadan itibaren giderek artan dirençle iş-uğraşı tedavisine geçilirken, 4. aydan itibaren hasta günlük yaşamda elini kullanmada tamamen serbest bırakılmıştır. Cerrahi sonrası 6., 8. ve 12. haftalarda yapılan değerlendirmelerde başparmakların eklem hareket açıklığı, kavrama ve çimdikleyici kuvvetlerinde belirgin iyileşme olduğu görülmüş; bu sonuçlar elin fonksiyonel seviyesine de olumlu yönde yansımıştır. Hastamızda okul çağından önce başparmak ekstansiyonunun cerrahi ve el rehabilitasyonu ile restore edilmesi el fonksiyonu ve yaşam kalitesini optimal seviyeye çıkarmıştır.

Anahtar kelimeler: *Başparmak hipoplazisi, el fonksiyonu, el terapisi*

Thumb hypoplasia and radial dysplasia among children are the conditions most frequently referred to hand therapy. There are various forms of thumb hypoplasia, which occur most commonly as part of radial deficiency. The underdeveloped thumb has been classified into five types to guide treatment recommendations (Table 1) [(1)]. Type II deficiency is characterized by thumb-index web space narrowing, absence of the thenar muscles and instability of the metacarpophalengeal joint of

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the thumb. Type III hypoplasia includes the intrinsic anomalies associated with a type II deformity, and additional skeletal and extrinsic musculotendinous abnormalities. Many descriptions have been made about the congenital absence of thenar muscles, thumb intrinsic and extrinsic muscles and their management and rehabilitation [2-4]. Since then, no cases with congenital absence of bilateral extensor pollicis longus (EPL) and extensor pollicis brevis (EPB) and its management and rehabilitation of the thumb function have been reported.

Table 1. Classification of hypoplastic thumb.

Type	Clinical Features
I	Gross size diminished
II	Narrow first web space; hypoplastic thenar muscles; MCP joint instability
III	Narrow first web space; hypoplastic thenar muscles; MCP joint instability; abnormal extrinsic tendons; hypoplastic metacarpal; IIIA with stable CMC joint; IIIB with unstable CMC joint
IV	Pouce flottant (floating thumb); rudimentary phalanges; skin bridge with neurovascular pedicle
V	Absent thumb

MCP: metacarpophalangeal, CMC: carpometacarpal

Achieving optimal upper limb function is of priority in the treatment of children with congenital hand anomalies [5]. The thumb plays a key component in hand function. This case reports the restoration of thumb extensor function with a detailed physiotherapy program in a child with bilateral congenital EPL and EPB absences.

CASE REPORT

Our patient was a 5 year-old girl and only child of the family. Abnormal motion pattern and position of the thumb, and dropped thumb, were noticed by the family when she was 3 years old (Figure 1). Clinical evaluation by the orthopeadic surgeon detected bilateral absence of thumb interphalangeal (IP) and carpometacarpal (CMC) extension and this finding was supported by the magnetic resonance imaging. Certain upper extremity anomalies are isolated cases not related to other musculoskeletal problems, whereas others are associated with systemic conditions [6]. One of the most common anomalies associated with systemic conditions is hypoplastic thumb [7]. To eliminate systemic problems, genetic and renal defects, chromosomal analysis and renal ultrasound imaging were performed. Chromosomal analysis resulted in a 46-XX genotype, and renal ultrasound imaging was unremarkable.

Figure 1. Dropped thumb before the surgery.

The girl underwent transfer of lateral extensor indicis proprius to extensor pollicis longus tendon and immobilization for 6 weeks wearing a forearm cast with 20° wrist extension and thumb in full extension and abduction [8]. After removal of the conventional forearm cast and Kirschner wires, rehabilitation program was initiated. With early dynamic motion protocol wearing a forearm splint with limited but progressively increased active flexion of the IP joint was combined with passive extension through a wire-rubber band system. The rubber band system was connected to a ermoplastic splint that had 20° wrist extension, full thumb extension and abduction (Figure 2). The splint was removed by the hand therapist to enable active and assisted flexion and active full extension of thumb IP joint in rehabilitation sessions. By removal of the splint, therapeutic ultrasound was applied to facilitate healing of the tendon and connective tissue during the rehabilitation sessions 5 days of the week for 2 weeks. Between 5th and 8th weeks, the protocol consisted of assisted active flexion and extension, movement against increased resistance and passive motion of the joints that have not reached full range of motion after removal of the splint. After 8 weeks, occupational task with increased stress loading and the challenging daily activities for the hand were added to the program through the 12.

week. After 4 months, the child was allowed to return to daily life activities.

Figure 2. Dynamic splint with a wire-rubber band system.

Standardized follow-up evaluations were performed at 6th, 8th and 12th weeks after surgery. Active ROM of the thumb MCP and IP joints were measured with a goniometer. Grip strengths were measured with a computer-based Jamar dynamometer. Pinch grip was measured with an electronic pinchmeter. Hand dexterity was determined by 9-Hole Peg Test (9HPT) [9].

Assessment of thumb IP and MCP range of motion was performed preoperatively and at postoperative 6th, 8th and 12th weeks. Although there was no active flexion and extension in MCP and IP joints preoperatively, an improvement was seen in extention of both MCP and IP joints in post-operative assessments (Table 2).

Table 2. The results of range of motion in both hands pre-operative and post-operative.

	RIGHT				LEFT			
Assessments	**Pre-op**	**1st**	**2nd**	**3rd**	**Pre-op**	**1st**	**2nd**	**3rd**
MCP flexion	90°	20°	50°	50°	90°	15°	50°	50°
MCP extension limitation	50°	15°	0	0	50°	10°	0	0
IP flexion	80°	30°	80°	80°	80°	25°	70°	80°
IP extension limitation	80°	10°	0	0	80°	10°	0	0

1st, 2nd and 3rd assessments (MCP:metacarpophalangeal joint; IP: interphalangeal joint).

Table 3. The performance results of 9 Hole Peg Test in the post-operative.

	RIGHT		LEFT	
Assessments	**Place (sec)**	**Remove (sec)**	**Place (sec)**	**Remove (sec)**
1st	70.2	50.3	71.3	52.1
2nd	60.3	30.3	62.7	29.8
3rd	45.2		45.5	

1st, 2nd and 3rd assessments.

The 9 HPT performance improved in both hands in the post-operative assessments with a significant difference in the last assessment (Table 3).

Gross grasping and pinching strength results are shown in Figure 3. It is seen that gross grasping strength of both hands increased briefly with treatment. Pinch strengths also improved by the time with a dramatic rise especially in lateral and triple pinchs in both hands.

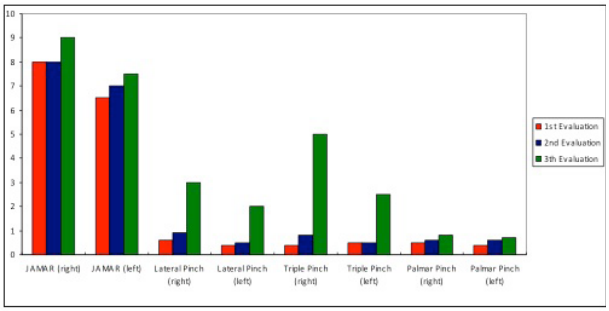

Figure 3. The results of gross grasping and pinching strengths in both hands comparing the post-operative 1st, 2nd and 3rd evaluations.

DISCUSSION

Congenital anomalies like thumb hypoplasia and radial dysplasia among children are the conditions frequently referred to hand therapy. The challenge of evaluating the function of children with congenital hand anomalies is to provide a detailed assessment about musculoskeletal composition of the child that has an impact on hand function. This is necessary for planning and success of the surgery and implementing effective physiotherapy programs.

Children with such conditions require extensive functional assessments for the success of surgical procedure and rehabilitation program. The thumb plays a key role in hand functions. Already in utero and thus before being able to grasp objects voluntarily, infants use the thumb sucking for spontaneous movements [10,11]. In addition, thumb sucking in utero or for spontaneous movements at birth are good predictors of future handedness and hand

functions [12,13]. Verdan reports that hand functions reduced by % 40 if the thumb is not present [14]. Its rotation in an opposing position against the remaining digits in the hand is a necessary component of almost all gross and fine grasps of the hand in daily activities [15]. Although thenar muscles produce the opposition of the thumb, EPL helps this motion by extending second phalanx and cause retroposition of metacarpals. By extending MCP joint when the IP joint is flexed, the EPB plays an important role in the precision grip and pinch. Especially the function of the thumb and index finger in grasping the cylindrical objects are very important [16]. Restorative surgery supported with a well-structured physiotherapy program improved the range of flexion and extension motions of the thumb within 12 weeks. Increase in the strength of that gained range of motion brought out more powerful hand grip and pinch. Acquisitions through such a short period are important for the child's future life.

Using hand and thumb in daily activities by grip and pinching makes humans different from the other animals. Inability to use the hand in daily activities because of the congenital anomalies and orthopedic impairments cause disability in the chronic stages. Ing and Tewey estimated that the percentage of the children with disability ranges from 2.6 % in children 0-3 years of age up to 12.4 % in children 15-17 years of age [17]. Expectations about functional status of the children with congenital anomalies change through maturation. Thus, the decision for surgery should be made when the child is at a younger age, before school age and prior to developing interests and skills.

Restoration of the thumb extension should be performed urgently, just like in this case with bilateral absence of thumb extensors. By this way, functional status improved and disability level reduced in the early stages of this child's life.

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