The Result of Correction Osteotomy of the Radius and Shortening Osteotomy of the Ulna for Madelung Deformity: Case series

Madelung Deformitesi Tedavisinde Radius Düzeltici, Ulna Kısaltıcı Osteotomi Sonuçları: Olgu serisi

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Abstract

Aim: Madelung deformity is a rare condition characterized by premature growth arrest of the volar–ulnar aspect of the distal radius growth plate. Different surgical procedures are described in the literature. In this study, we aimed to present functional and clinical results of our technique in five wrists of 4 patients.

Materials and Methods: Five wrists of 4 patients (3 females and 1 male) underwent surgery for Madelung deformity in our clinic between 2005 and 2014. The mean age of the patients was 15.4 (14–17) and the mean follow up duration was 65.6 months (29–111). One patient had bilateral deformity. The patients were evaluated in terms of wrist range of motion (ROM), DASH score, and VAS score pre-and post-operatively. Corrective osteotomy to the radius, shortening osteotomy to the ulna, and release of the Vickers ligament were performed on all of the patients.

Results: Although there were no meaningful changes in the range of supination–pronation, palmar flexion was decreased, and dorsiflexion and radial deviation were significantly higher in the last follow-up compared to the preoperative values. Union was achieved at all of the osteotomy sites. All radiological parameters were significantly better in the last follow-up X-rays. The preoperative and postoperative mean DASH scores were 55.4 and 18.8, respectively, and the preoperative and postoperative mean VAS scores were 6 and 1, respectively.

Discussion and Conclusion: This surgical technique may be a good option for severe Madelung deformity in terms of functional and radiological results and patient satisfaction in the midterm follow-up. Pain and gross deformity were our main surgical indications.

Keywords: Madelung deformity; osteotomy; wrist; radius; ulna.

Öz

Amaç: Madelung deformitesi, radius distal büyüme plağının volar ve ulnar tarafında erken büyüme duraklaması ile tanımlanan nadir bir deformitedir. Tedavisi için literatürde farklı cerrahi girişimler tanımlanmıştır. Bu çalışmada Madelung deformitesinde kullandığımız cerrahi tekniğin klinik ve radyolojik sonuçlarını değerlendirmeyi amaçladık.

Gereç ve Yöntemler: 2005–2014 yılları arasında kliniğimizde 4 hastaya (3 kadın, 1 erkek) ait beş el bileği Madelung deformitesi nedeniyle opere edildi. Hastaların ortalama yaşı 15,4 (14–17) ve ortalama takip süresi 65 (29–111) ay idi. Bir hastada bilateral deformite vardı. Hastalar ameliyat öncesi ve sonrası eklem hareket açıklığı, DASH ve VAS skorları ile değerlendirildi. Tüm hastalara radius düzeltici osteotomi, ulna kısaltıcı osteotomi ve Vickers bağı gevşetme ameliyatı uygulandı.

Bulgular: Hastaların son kontrollerindeki eklem hareket açıklıkları ameliyat öncesi değerler ile karşılaştırıldığında, dorsifleksiyon ve radyal deviasyon aralığı anlamlı olarak artarken palmar fleksiyon arkı azalmıştı. Supinasyon ve pronasyon aralığında anlamlı bir değişiklik izlenmedi. Tüm osteotomi alanlarında kaynama sağlanmıştı. Son takip röntgenlerinde tüm radyolojik parametreler ameliyat öncesine göre anlamlı olarak daha iyiydi. Ameliyat öncesi ve sonrası ortalama DASH skoru sırasıyla 55,4 ve 18,8 iken, ameliyat öncesi ve sonrası VAS skoru sırasıyla ortalama 6 ve 1 idi.

Tartışma ve Sonuç: İleri Madelung deformitesi olan hastalarda bu cerrahi teknik hasta memnuniyeti, fonksiyonel ve radyolojik sonuçlar bakımından iyi bir seçenek olabilir. Ağrı ve belirgin deformite bizim ana cerrahi endikasyonumuz oldu.

Anahtar Sözcükler: Madelung deformitesi; osteotomi; el bileği; radius; ulna

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INTRODUCTION

Madelung deformity was first described by the German surgeon Otto Wilhelm Madelung with the definition of deformity during a female autopsy (1). Later many publications mentioned this deformity.

Madelung deformity is a rare condition characterized by the premature growth arrest of the volar–ulnar aspect of the distal radius growth plate, causing pain and loss of function. Clinically soft tissue pathologies secondary to bony deformities and vascular digenesis may be seen.

In this deformity, growth of the unaffected parts of radial growth plate causes bending of distal radius to the volar and ulnar side. With extensive deformity, separation of the radio ulnar joint, dorsal protuberance of the ulnar head, and lunate deformity are seen. These anatomical changes result especially in limited extension and radial deviation, decreased grasping power, and often pain due to disrupting wrist biomechanics (2, 3) (Figure 1).

The deformity is seen more often in females, usually in late childhood and adolescence. (F/M: 4/1). It is bilateral in 50% of the cases and autosomal dominant in one third. Various publications report that Madelung deformity might be associated with multiple hereditary osteochondromatosis, Ollier disease, achondroplasia, multiple epiphyseal dysplasia, Turner syndrome and mucopolysaccharidosis. Additionally, it has been shown that this deformity is strongly associated with Léri-Weill dyschondrosteosis (4–7).

While its etiology has not been completely understood, genetic factors are thought to be present at its background. In addition, patients' nutrition status, occupation, vascular insufficiency at the distal growth plate, history of muscular diseases, trauma and infection with accompanied disorganization of the cells at the growth plate might play a role in the development of the deformity (2,6).

Asymptomatic patients can be followed conservatively; however, conservative treatment fails in preventing deformity development. There are many surgical procedures for the treatment of Madelung deformity (6–13). An appropriate procedure can be decided by taking into consideration the patient's age, degree of deformity and pain, and functional status. Osteotomy techniques such as dome osteotomy and transvers osteotomy have been defined in the literature (14,15). In our technique, we used parallel osteotomy to the deformed joint line. An open wedge operation to restore the length of the radius was performed for acute correction. Fixation can be obtained by external fixation, K-wires or plate fixation. We preferred plate fixation for rigid fixation (Figure 2). In the literature, Darrach or Sauve–Kapandji osteotomies are described for the ulna. In our series, considering the wrist biomechanics, we performed ulnar shortening and osteosynthesis.

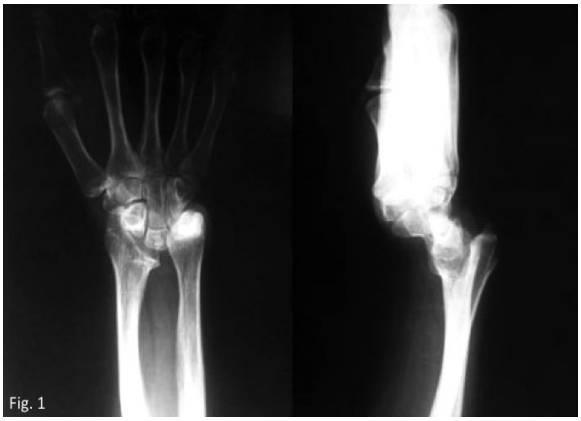

Figure 1. Pre-op wrist AP and lateral X-ray of the patient with Madelung deformity. Note the classical features of the deformity, such as the bending of the distal radius to the volar and ulnar side, separation of radio ulnar joint, dorsal protuberance of the ulnar head, and deformity of the lunate bone.

Severe deformity and pain were the chief complaints in all patients who underwent surgery in our clinic, a tertiary care center. In all of the patients, corrective osteotomy to the radius, shortening osteotomy to the ulna, and releasing of the Vickers ligament were performed. In this study, we aimed to present functional and clinical results of this technique using the data on five operated wrists.

MATERIAL AND METHODS

After obtaining the approval of our institution's institutional review board, patient charts were reviewed retrospectively. Written informed consent was obtained before beginning the treatment. Five wrists of 4 patients (3 females, 1 male) were operated for Madelung deformity in our clinic between 2005 and 2014. The mean age of the patients was 15.4 (14–17) and the mean follow up duration was 65.6 months (29–111).

Table 1. Demographical and clinical features of the patients

					Dorsiflexion (°) (P=0.042)		Palmar flexion (°) (P=0.039)		Radial deviation (°) (P =0.025)		Supination–pronation range (°) (P>0.05)		DASH (P = 0.042)		VAS (P = 0.043)	
No	**Sex**	**Age at op.**	**Side**	**Follow-up**	**Pre-op**	**Post-op**	**Pre-op**	**Post-op**	**Pre-op**	**Post-op**	**Pre-op**	**Post-op**	**Pre-op**	**Post-op**	**Pre-op**	**Post-op**
1	M	14	R	103	25	45	80	76	25	35	140	130	43	24	4	0
2	F	16	L	111	60	65	70	68	15	25	160	140	43	29	6	1
3	F	14	L	55	40	75	75	72	10	20	145	135	62	14	5	2
4	F	16	R	30	40	85	72	70	15	25	145	149	63	9	8	1
5	F	17	R	29	60	80	70	68	10	20	150	150	66	18	7	1
M		15.4		65.6	45	70	73.4	70.8	15	25	148	141	55.4	18.8	6	1

DASH: The Disabilities of the Arm, Shoulder and Hand ScoreVAS: Visual Analogue Scale
M: Mean

Table 2. Pre- and post-operative radiological measurements of the patients

Wrist no	**Radial inclination (°) (P=0.039)**		**Ulnar variant (9) (P = 0.043)**		**Subluxation of the ulna (9) P=0.043**		**Volar tilt (°) (P = 0.042)**	
	Pre-op	**Post-op**	**Pre-op**	**Post-op**	**Pre-op**	**Post-op**	**Pre-op**	**Post-op**
1	50	15	28	-4	16	8	30	22
2	34	21	4	-2	13	8	16	11
3	60	25	15	0	10	4	18	12
4	60	20	15	2	14	2	20	16
5	60	25	18	-6	16	2	25	20
Mean	52.8	21.2	16	-2	13.8	4.8	21.8	16.2

One patient had bilateral deformity. Patients were evaluated in terms of wrist ROM, DASH score, and VAS score pre- and post-operatively. Patients' ROM were assessed with the patients sitting upright and their elbows at 90% flexion and forearms at supination. Ulnar variance, radial inclination angle, volar tilt and dorsal subluxation degree of the ulna were evaluated on the X-rays. Dorsal subluxation of the ulna was measured as the distance between the radius and dorsal cortex of the ulna on the lateral wrist X-ray (16). Patients did not have any associated genetic disorders. The chief complaint was deformity and pain in all patients.

SURGICAL TECHNIQUE

AP and lateral X-rays were taken before surgery for radiological evaluation. Osteotomies and bone resections were planned on the basis of ideal values (distal radial inclination: 22–23 degrees at AP view, volar tilt: 10–11 degrees on lateral view, and ulnar variance: 0

degrees) (Figure 3). Templates were used to see the final position. Volar approach was used to cut the Vickers ligament. Then, pronator quadratus was dissected from the radial side of the muscle between flexor carpi radialis and radial artery. Open wedge osteotomy was done parallel to the joint line under fluoroscopy. Correction level was determined by using fluoroscopy and in accordance with the previously mentioned ideal values. Shortening osteotomy of the ulna was performed by an incision over the distal ulna. Plate and screws were used for the fixation of the ulna and radius in 4 wrists. Kirschner and cerclage wire were used for the other.

The resected ulna segment was used as a structural autograft at the radial osteotomy site with appropriate shape in three wrists. Thus, additional donor site morbidity was not caused and positive ulnar variance was corrected. None of the patients had any complications due to the surgery. Patients were followed by cast immobilization until the union was obtained. Union was seen on the follow-up X-rays to have been achieved in all of the patients after a mean postoperative duration of 6 weeks (Figure 4). Physical therapy was started after the removal of the cast to gain maximal functionality.

STATISTICAL ANALYSIS

The differences in pre-op and last follow-up values of the parameters were analyzed by using Wilcoxon test. $p<0.05$ was accepted as significant.

RESULTS

When the mean pre-op and post-op clinical data were analyzed, the values for dorsiflexion were 45–70, palmar flexion 73–70, radial deviation 15–25, and range of the supination-pronation 148–139, respectively. The mean preoperative DASH and VAS scores was 55.4 and 6, respectively. Postoperatively these scores were 18.8 and 1, respectively. Although the change in the range of supination–pronation was not significant between the pre-operative and last follow-up period, palmar flexion was seen to have decreased in the last follow-up examination. Other clinical parameters were significantly higher in the last follow-up visit, compared to the preoperative values (Table 1).

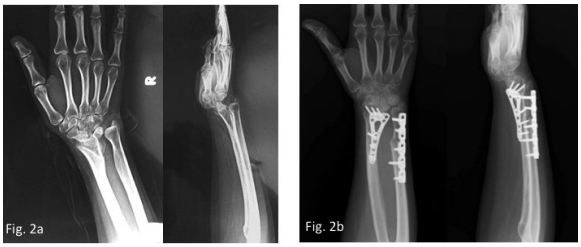

Figure 2. Preoperative AP and lateral wrist X-rays of the 16 year-old-female patient with Madelung deformity (Figure 2a). Postoperative AP and lateral wrist X-rays of the same patient in month 24 (Figure 2b).

In the radiological evaluation, pre- and post-operative mean radial inclination angle was 52.8–21.2 degrees, mean ulnar variance 16 – minus 2 mm, mean dorsal subluxation of the ulnar head 13.8–4.8 mm, and volar tilt 21.8–16.2 degrees, respectively. Union was achieved in all of the osteotomy sites. All radiological parameters were read as significantly better in the last follow-up X-rays (Table 2).

DISCUSSION

Although Madelung deformity is rarely seen, it comprises 1.7% of congenital deformities (17). Although this deformity is associated with various genetic diseases, vascular insufficiency at the distal growth plate, muscular diseases, disorganization at the growth plate caused by trauma and infection, and the patient's nutrition status and occupation might also play a role in its development (2,6). The classification of the deformity is generally based on the etiology in the literature; but there is no established classification system in practice.

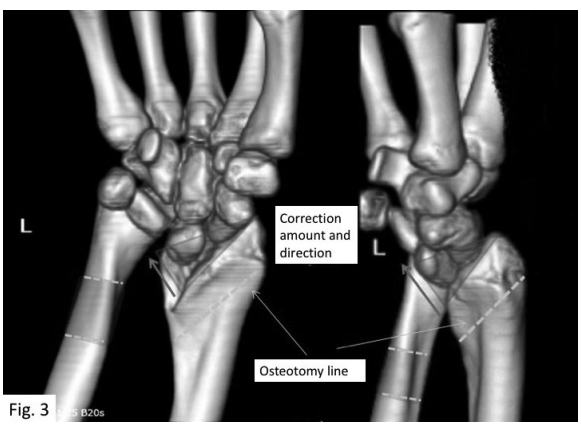

Figure 3. Surgical technique shown on reconstructed CT image. Osteotomy in parallel to joint line shown as dashed yellow line and targeted correction angle as triangle.

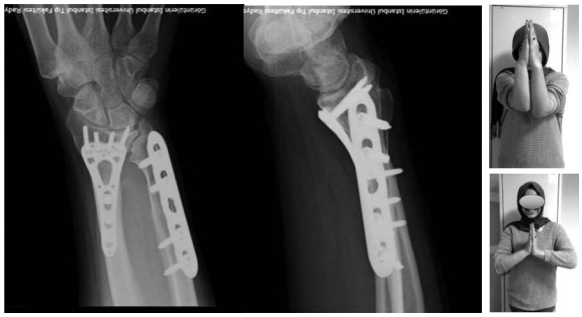

Figure 4. AP and lateral wrist X-rays and clinical images of the same patient in postoperative month 17.

In clinical practice, lateral radiographs of the wrist are usually taken by aligning shafts of the ulna and radius. While it serves the purpose in most cases, in cases such as ours where the relation of the radius and ulna may be disrupted true lateral radiographs defined according to carpal bones are warranted (18).

All patients in our series had deformities accompanied by pain and loss of function. None of them had any associated genetic disorders.

As a result of the anatomical changes in Madelung deformity, wrist biomechanics is disturbed, grasping power and ROM is decreased. Main objective of the surgery is to restore normal anatomy as much as possible. There are no universally accepted guidelines to decide when to operate. Many factors such as functional and anatomic changes, cosmetic concerns, associated nerve compressions, extensor tendon ruptures can be indications for surgery (7,9–13,19). However, complete correction of this deformity can result in instability because of the impairment of the balance between triangular complex and joint facets in patients with severe deformity. Thus, partial correction might be more acceptable in such patients.

Surgical treatment options include soft tissue procedures (Vickers ligament release) epiphysiodesis, ulna shortening procedures (Darrach or Sauve–Kapandji osteotomies), combination of distal radius correction and ulna shortening osteotomy or radius dome osteotomy. (6,7,9–14,20).

In the study of Steinman et al., good and excellent results were reported in the 11 years' follow-up of nineteen patients who had undergone volar ligament release and radius dome osteotomy. They emphasized that tri-planar deformity could be corrected more consistently with dome osteotomy (14). Transverse osteotomy to the radius and deformity correction and lengthening by use of Ilizarov method are also suggested by authors in the literature (15). It was also shown by Baskır et al. from our clinic that good results could be achieved by using Darrach procedure in nine cases (20). We preferred Vickers ligament release, distal radius open wedge osteotomy, ulnar shortening osteotomy and autograft with ulna segment in all of the patients, and obtained successful results by this technique.

Correction of volar tilt is important for wrist dorsiflexion, grasping power and appearance (6). In this series, a significant correction has been obtained in the radial inclination angle, ulnar variance, volar tilt and ulnar dorsal subluxation values. This radiological correction is also reflected in the ROM of the wrist joint as a significant improvement in wrist dorsiflexion and radial deviation degrees.

Although there is no consensus about the timing of the surgery, it is recommended that bone procedures such as correcting osteotomies and shortening should be performed after the closing of the physis to avoid recurrent deformities (6). Ulnar head resection at early childhood can result in ulnar deviation and instability. Based on our experience, surgery is more suitable for severe deformity and should be performed after the closing of the growth plates. Mild and moderate deformities can be treated without surgery.

Although we lack long-term follow-up results, complications such as infection, reflex sympathetic dystrophy and constant pain are reported in the literature. Schmidt-Rohlfingin et al. reported a case followed up for 25 years without surgery, with radiological arthritic changes in the wrist (12). In our series with a mean duration of 65.6 months, no complication was observed during the follow-up.

The main reason of the pain is the thickened radiolunate ligament, which is called the Vickers ligament in patients with incomplete bone growth. In Madelung deformity radiolunate ligament is thickened and connects the lunate bone and triangular fibrocartilage complex to the deformed radius. Release of this structure with or without osteotomies can be helpful for deformity correction and pain relief (9). It was released in all of our patients.

During the pre-operative planning of the multiplane osteotomies in Madelung deformity, computerized modeling can be useful for performing patient-specific osteotomies. However, soft tissue releases should be taken into account as well and the most comfortable approach should be chosen. Based on the limited experience we have acquired, we believe that union will not be problematic when we consider microcirculations of the distal radius. Thus, we think that ulnar autograft can be used without union problems. Our series supports this idea, given that union was achieved in all of the osteotomies.

The main limitation of this study is the relatively small number of patients; however, since the deformity is rarely seen, multicenter studies are required to obtain an adequate number of patients. It can be regarded as another limitation that we could not use the grasping strength as a parameter.

CONCLUSION

In conclusion, this surgical technique may be a good surgical option to treat severe Madelung deformity with respect to functional and radiological results and patient satisfaction in the midterm follow-up. Pain and severe deformity were the main indications for surgery in our series. Further studies are needed for a better understanding of the potential of this surgical technique in the treatment of Madelung deformity.

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