Correction of Madelung deformity by monorail external fixator in young adults

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Technical advance

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Abstract

Aims: Many surgical techniques are proposed to address Madelung deformity, but the most suitable surgical treatment still remains unknown for symptomatic cases. We introduced a modified management by radius osteotomy and Sauvé-Kapandji's procedure using the monorail external fixator to provide a supplement to the treatment of Madelung deformity.

Methods : From January 2013 to October 2018, 12 patients (9 right and 6 left wrists) with a mean age of 18 (16 to 24) years complained of pain, deformities and limited function of effected wrists. These 12 patients received operation for subsequent acute correction by monorail external fixator. The Vas Score, ulnar tilt of the radial articular surface angle, radiopalmar tilt angle, lunate subsidence, volar translation and range of wrist movement were evaluated pre- and postoperatively.

Results : Consolidation cost an average time of 87 (66 to 122) days. All patients were followed up for an average time of 24 (8 to 65) months. The Vas score for all patients decreased from 4 (2 to 7) to 0 ($p<0.001$). The ulnar tilt and radiopalmar tilt were decreased by a mean of 21° (mean changed from 50 to 29) and 21° (from 38 to 17), respectively (both $p<0.001$). The length of lunate subsidence and volar translation was reduced by a mean of 6 (from 7 to 1) mm and 8 (from 25 to 17) mm, respectively (both $p<0.001$). Five patients received radius lengthening for an average of 22 (18 to 29) mm. Wrist extension improved by a mean of 8° (from 50 to 58), while flexion decreased by 40° (from 100 to 40).

Conclusion : Acute correction by radius osteotomy and Sauvé-Kapandji's procedure and gradual lengthening with the extendable monorail external fixator can be used for Madelung deformity with little surgical trauma. The radiographic parameters and wrist pain are improved after surgery and postoperative therapy.

Introduction

In 1878, Otto W. Madelung first provided an overview of a rare disease of the wrist, now known as Madelung deformity (MD) [1], and described a palmar subluxation of the hand, a prominent distal ulna, and volar angulation of the distal radius epiphysis, even before the discovery of radiographs. MD often occurs bilaterally but is unequally affected, with onset during adolescence [2]. Through many studies of radiographs and pathology, we now know the deformity caused by the premature partial closure of the ulnar part of the distal growth plate of the radius [3]. Volar and medial epiphysiodesis of the distal radius results in the forward tilt of the radius combined with the posterior displacement of the ulna, which produces an anterior displacement of the wrist. Patients often complained of wrist pain, visible deformities in the forearms and limited range of movement (ROM) [4].

Nonoperative management is ineffective in treating persistently painful wrists and does not prevent the progression of the deformity. Therefore, surgery is recommended to treat MD. The aim of surgical treatments is mainly to correct the position of the distal radial articular surface and to restore satisfactory radial and ulnar anatomy. Surgical therapies can be roughly classified into three major groups: (1) a procedure of the radius, such as correction osteotomy, epiphysiodesis, or progressive lengthening[5,6,7,8]; (2) a procedure of the ulna, including shortening osteotomy and head excision [9,10,11]; and (3) a combined procedure of both bones, such as Sauvé-Kapandji's procedure or ulnar shortening combined with radial osteotomy [10,12,13]. Recent procedures have preferred to deal with the ulna and radius at the same time [10,14,15] because complications or secondary operations, such as ulnar subluxation of the carpus or radius lengthening after operation, have occurred in isolated ulna or radius surgeries.

The combined procedure of both bones brought more normal wrist biomechanics and more invasions. We proposed modified management using a monorail external fixator technique to decrease trauma to the operation. This monorail external fixator is equipped with Micrometric Swiveling Clamps (MSCs), and the three-dimensional deformity of the distal forearm can be corrected through MSCs after Sauvé-Kapandji procedure and radius osteotomy are performed. This management included the restoration of the distal radioulnar joint (DRUJ), improvement of ROM, and potential lengthening of the forearm.

Patients And Methods

Patients

From January 2013 to October 2018, 12 patients [10 females and 2 males, 9 right and 6 left wrists, with a mean age of 18 (16 to 24) years at operation] in this series were all of primary origin, and the skeleton was mature at the time of surgery. Pain and deformity were the presenting features in all patients. The patient details are summarized in Table 1 and Table 2.

All patient clinical features were assessed by the authors. A visual analog score (VAS, 0~10) was used to evaluate the pain before the operation. Range of motion (pronation/supination, flexion/extension) and radiographic data (ulnar tilt of radial articular surface, radiopalmar tilt, lunate subsidence and volar translation) were measured preoperatively [3,16]. Because the radius was bowed, the longitudinal and transverse axes could not be determined. The ulna was usually straight, providing a more reliable longitudinal axis [17]. Preoperative clinical data are shown in Table 1 and Table 3.

Surgical technique

Surgeries were performed under brachial plexus block or general endotracheal anesthesia based on patient risk factors and preference. The affected limbs were exposed after disinfection. The first Schanz screw was drilled into the distal end of the radius below the articular surface through the skin in the coronal plane under X-ray fluoroscopy. The first Schanz screw was installed at the distal end of MSCs, and the fixator rail was maintained parallel to the radial shaft in the coronal plane. The second screw for the fixator was inserted proximally to the radius shaft through the proximal clamp of the fixator as a proximal reference. Under lateral wrist X-ray fluoroscopy, the third screw entry point was predesigned. A line passed through the entry point of the first screw at an angle of α to the tangent line of the radial articular surface (α was approximately 70–80 degrees because the tangent line of the radial articular surface and axis of the radius shaft were at an angle of 70–80 degrees in the sagittal plane for normal wrists). The entry point was located on the line at the position 2–3 cm proximal from the first screw. The third and fourth screws (near the first screw) were drilled into the radius, and all screws, except the third screw, were installed into the frame to make sure that the frame was stable. Vertical radius osteotomy and ulna tilt were performed through the radialis side of the radius and the ulnar side of 1 cm incisions at the distal metaphysis by electric drill and osteotome. The MSC was first adjusted in the coronal plane to correct the ulnar tilt of the radial articular surface, and then the third screw and other screws in the rail were aligned to correct the radiopalmar tilt. Then, the Sauvé-Kapandji procedure was performed in the same steps as described by Angelini et al. to increase the stability of the new carpus and improve the rotation function of the forearm [12]. K-wires were used for temporary fixation and then the remaining screws were drilled along the rail. After completing angular correction with the MSC, X-rays were taken to confirm satisfactory angular correction and screw placement. Finally, the wounds were closed. The procedure of correction is shown in Fig 1.

Postoperation

The tissue adjacent to the pins was compressed with gauze dressings to keep the wound dry and prevent pin tract infections. The patients were told to start functional exercise (clenching fists and extending hands) at an early stage to prevent the adhesion of wrist tendons. Operated limbs were permitted no load for 6 weeks. If a callus appeared, then the load of the operated limbs was gradually increased to promote osteogenesis. Patients with shortening forearms were told to start lengthening at 10 days after operation at a speed of 0.75 mm per day until their forearms were sufficiently lengthened. The frame and screws were removed after fracture healing was completed. At the first follow-up after frame removal, the clinical features were assessed again. Nonparametric sign tests were used to test for differences between preoperative and postoperative measurements. Postoperative clinical measurements are shown in Table 3.

Results

All patients were followed up after the operation with a mean duration of 24 (8 to 65) months. An average time of 87 (66 to 122) days for consolidation was completed postoperatively. All patients were free from pain, with an average pain score(Visual Analogue Score, VAS) change from 4 (2–7) before the operation to 0 at the first follow-up after frame removal ($p<0.001$).

The ulnar tilt of the radial articular surface, radiopalmar tilt, and lunate subsidence volar translation were mainly decreased to the normal range postoperatively. The ulnar tilt of the radial articular surface and radiopalmar tilt were decreased by a mean of 21° (from 50° to 29°) and 21° (from 38° to 17°), respectively (both $p<0.001$). The length of lunate subsidence and volar translation were reduced by a mean of 6 (from 7 to 1) mm and 8 (from 25 to 17) mm, respectively (both $p<0.001$). Five patients received radius lengthening for an average of 22 (18 to 29) mm. A 24-year-old woman (case 6) who suffered pain of right wrist for 5 years with her right forearm shortening about 2cm was treated by acute correction, and gradual lengthening with the monorail external fixator (Fig. 2).Wrist extension improved by a mean of 8°, while flexion decreased by 40°; supination of the forearm improved from 87° to 91°, and pronation increased from 50° to 55°. However, the improvement of rotation was not statistically significant. Range of wrist movement of an 18 year-old woman (case 11) showed the improvement of the operated wrist after eight months post-operative (Fig. 3).

Superficial pin tract infection occurred in one patient. The resulting local infection was controlled with wound care until the pins were removed. There were no deep pin-track infections, neurovascular injury nonunions or other complications. Otherwise, the patients tolerated the procedure well and were satisfied with the functional and cosmetic results.

Discussion

MD more commonly presents during the teenage years, and early surgery is usually indicated for pain and deformity. Of the several surgical techniques that have been proposed, our technique is the first to use the extendable monorail external fixator on wrists and adequately relieve pain and deformity.

Wrist pain is caused by persistent DRUJ incongruity. This pain is mainly due to excessive shear force on the radiocarpal joint secondary to the abnormal radioulnar tilt of the radial articular surface combined with ulnar impaction due to a relatively long ulna. Darrach and Nielsen et al. reported the excision of the distal ulna alone to treat the pain of MD [18,19]. The Darrach procedure completely eliminates the primary cause of pain but may leave the carpus unstable for further ulnar subluxation of the carpus. Consequently, the simple resection of the ulnar head has been largely abandoned. Bruno et al. reported ulnar-shortening osteotomy as a safe and reliable surgical procedure to relieve ulnar-sided wrist pain in 9 adult female patients with symptomatic MD [9] and reported that 2 of those patients experienced persistent pain after the operation. Ulnar-shortening osteotomy preserved wrist stability but left the radius deformity the same as observed preoperatively, which meant a risk of pain recurrence because the fundamental cause of pain remained unchanged.

The premature partial closure of the ulnar part of the distal growth plate of the radius leads to a bowed short radius and an excessively tilted radiocarpal joint surface. Surgeries of the radius are necessary for the correction of MD. Vickers et al. performed a resection of the ulnar zone of the distal radial epiphysis in 14 late-childhood patients to restore radius growth and prevent deformity [17], but 4 patients did not improve after the operation, and the improvement of the remaining patients was slight. The effect of Vickers' treatment is not significant, and this procedure can only be used in children whose epiphysis was open. The treatment of MD in children may not only be ineffective but also aggravate malformations, so some authors have proposed that correction operations should not be performed until the epiphysis is closed. Steinman et al. performed dome radius osteotomies for 19 adult patients with MD and obtained good improvement of deformity and ROM, but these authors also reported that 6 of those patients experienced persistent pain with recurrence [6]. Those 6 patients underwent repeat surgery for ulnar shortening (5 patients) or Sauvé-Kapandji's procedure (1 patient).

Moreover, recent authors have advocated addressing the ulna and radius at the same time, aiming to restore more normal biomechanics to the wrist. Kampa et al. performed radial opening wedge osteotomy combined with a modified Darrach procedure for 5 female patients and reported excellent outcomes in relieving pain and improving ROM [10]. However, the patients were all at a risk of carpus unstable. Laffosse JM corrected MD in 14 cases by combined radioulnar osteotomy, but there two patients still experienced pain during sustained activity [14]. Since DRUJ was not restored, it was possible that pain may reappear. White et al. reported a case of MD corrected by the Lauenstein procedure (the Sauvé-Kapandji procedure) and a closing wedge radius osteotomy [20]. The osteotomy of the radius improved the mechanics of the radiocarpal joint and prevented radiocarpal arthritis. Fusion of the distal radioulnar joint maintained carpal stability and relieved pain, and osteotomy of the ulna enhanced rotation. However, closing the wedge osteotomy further shortened the length of the bowed short radius. Therefore, a closing wedge radius osteotomy was undesirable. Another disadvantage of the combined operations was increased surgical invasions.

Houshian et al. [8] used the Ilizarov technique to gradually correct MD by isolated radius osteotomy in 7 patients and reported slight deformities and malalignments in the joints in some of the patients. The frame of the Ilizarov was large in size and inconvenient for daily life; furthermore, it was expensive to use the Ilizarov external fixator for treatment. Our management modified the extendable monorail external fixator with MSC to make it possible to correct the three-dimensional and shortening deformities of the radius at the same time as the Ilizarov external fixator, and the small volume reduced inconveniences and was less expensive. Another advantage of our management was microinvasiveness because screws and k-wires can be drilled through the skin with a tiny cut, there is no need for bone graft, and the pins can be removed without anesthesia.

Interestingly, the improvement of ROM was not statistically significant in our study. The range of wrist flexion in normal people was approximately 0 degree to 60 degrees, but that in patients with MD was approximately 0 degree to over 100 degrees. The excessive radiopalmar tilt of MD led to this change. In our study, the ulnar tilt of the radial articular surface and radiopalmar tilt were both corrected. The decrease in the radiopalmar tilt of the distal radius led to a decrease in the wrist flexion range, so we found that the flexion range of the wrist improved from too large to normal. The average pronation, supination, and extension of the wrists before and after the operation were improved but were not statistically significant. Except for the correction of bones, the role of the soft tissue, such as the ligament complex of the distal radioulnar joint and forearm interosseous membrane, in this situation may need to be further demonstrated.

Conclusions

The results of our study indicate that the monorail external fixator with the MSC system is able to achieve the correction of Madelung deformity, as measured radiographically, with less invasion. The monorail external fixator should be considered for the surgical treatment of Madelung deformity when the correction of a large angle and/or length is indicated.

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Tables

Table 1 Patient details

Variable	Number
Number of patients	12
Number of wrists	15
Male:female	10:2
Etiology(congenital:aquired)	12:0
Age in years at operation (mean; range)	18 (16-24)
Number of patients received lengthening	5
Total Consolidation time in days (mean; range)	87 (66-122)
Total follow-up time in months (mean; range)	24 (8-65)

Table 2 Percentage of patients' complaints

Complaints	Percentage
Pain	100(12)
Deformity of wrists	100(12)
Functional limitation of wrists	83.3(10)

Raw number in parentheses

Table 3 Correction of MD achieved by the MSCs system

Parameter	Preoperative	Postoperative	*P* value
Pronation(degree)	50(31-82)	55(33-78)	0.134
Supination(degree)	87(72-108)	91(78-100)	0.125
Flexion(degree)	100(92-108)	60(40-71)	0.001*
Extension(degree)	50(42-59)	58(34-79)	0.013*
Ulnar tilt(degree)	50(39-76)	29(20-47)	0.001*
Radiopalmar tilt(degree)	38(20-53)	17(13-20)	0.001*
Lunate subsidence(mm)	7(4-10)	1(-2 to 3)	0.001*
Volar translation(mm)	25(21-31)	17(14-20)	0.001*
VAS score	4(2-7)	0(0)	0.001*

Values are given as the mean (range)
* Statistically significant

Abbreviations

MD: Madelung deformity

ROM: range of movement

MSCs: Micrometric Swiveling Clamps

DRUJ: distal radioulnar joint

VAS: Visual Analogue Score

Declarations

Ethics approval and consent to participate

All patients receive written information among their participation in this study. They sign a written consent to participate in the study. The Ethics Committee of Shanghai Jiao Tong University Affiliated Sixth People's Hospital approved current study. All clinical investigations were performed in accordance with the guidelines of the Declaration of Helsinki.

Consent for publication

Not applicable.

Availability of data and materials

The datasets used and analysed in the current study are available from the corresponding author on reasonable request.

Competing interests

The authors have no relevant conflicts of interest to disclose in the preparation and completion of the manuscript.

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Authors' contributions

Li Shi: Date analysis, Wrote the manuscript; Meng-wei Wang: Data collection and analysis; Dong-dong Cheng: Data collection and analysis; Jia Xu: Data collection and analysis; Yimin Chai: Study design; Qinglin Kang: Performed surgeries, Edited manuscript.

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Figures

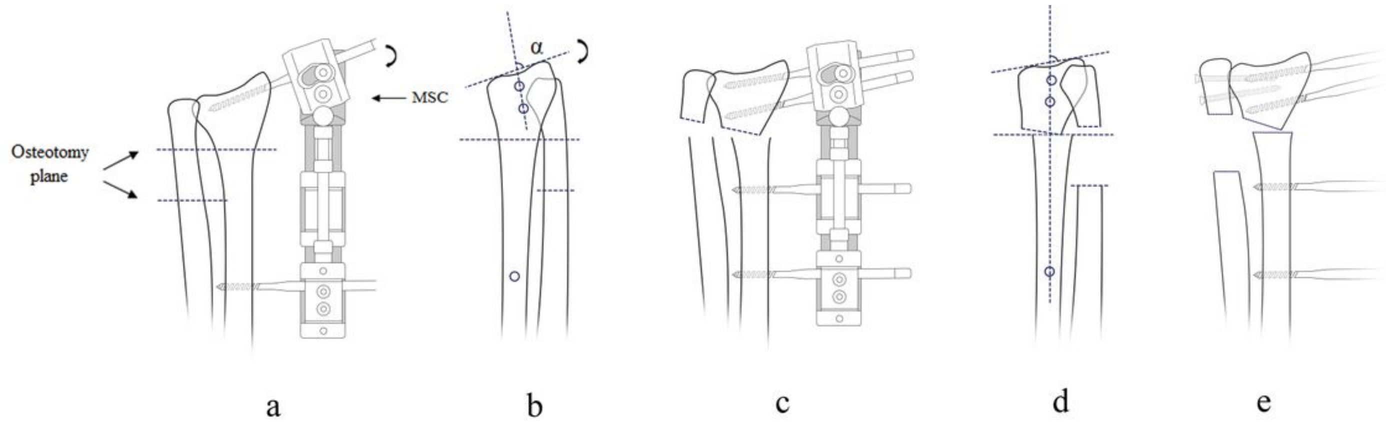

Figure 1

a) drill the distal and the proximal Schanz screws into radius in the coronal plane to install the external fixator; b) the angle α is pre-designed to position the third screw; c) install the third and the fourth screws to make sure frame stable and osteotomy at metaphysis of both ulna and radius then correct coronal plane deformity; d) correct sagittal deformity and perform Sauvé-Kapandji procedure; e) appearance after acute correction.

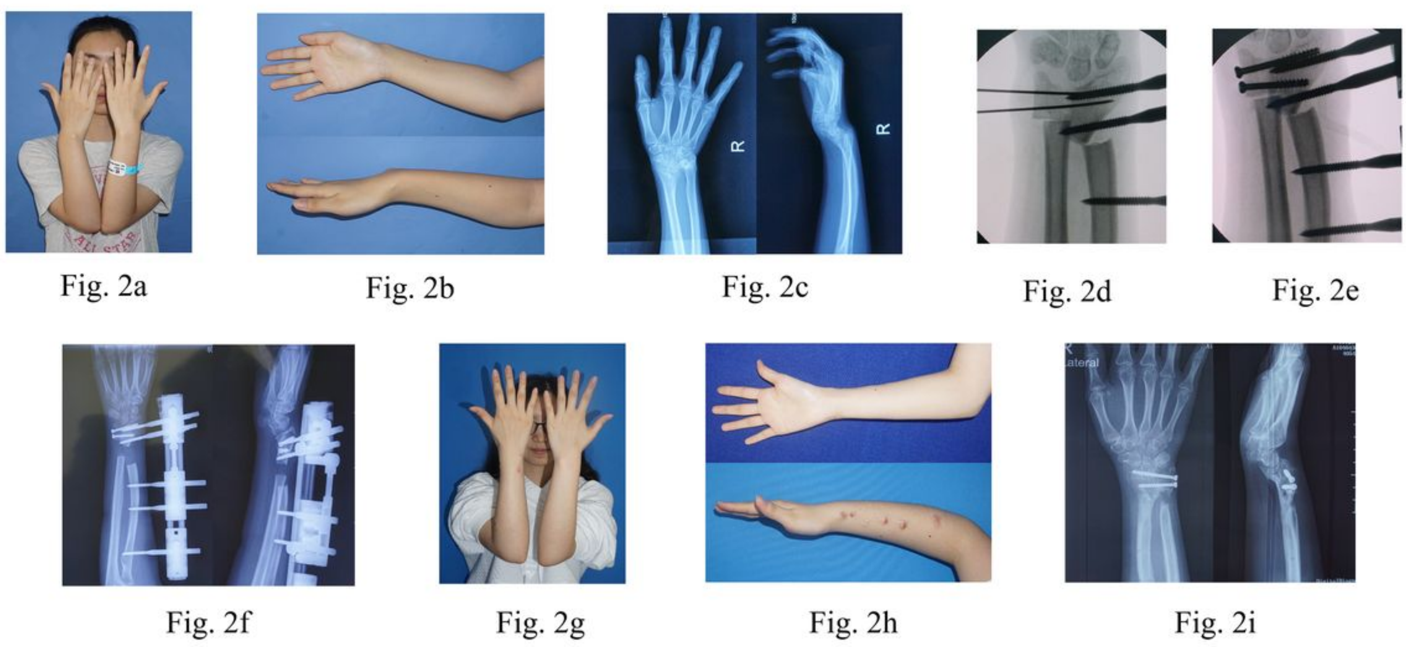

Figure 2

a) and b) a photograph; c) radiograph showing a palmar subluxation of the hand, a prominent distal ulna and volar angulation of the distal radius epiphysis; d) and e) fluoroscopic intra-operative views; f) post-operative view showing application of the monorail external fixator with a MSCs system to correct forearm shortening deformity; g) to i) photographs and radiograph at final follow-up postoperatively showing normal alignment and equal forearm lengths.

Figure 3

a) to d) photographs of wrist extension, flexion, pronation and supination before operation; e)to h) photographs of wrist extension, flexion, pronation and supination postoperative.