Original Article | Journal of Orthopaedic Case Reports 2026 July:16(7):Page 484-490

Outcomes of Surgical Treatment for Masada Type lVa Forearm Deformity in Children with Multiple Hereditary Exostosis: A Case Series

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Learning Point of the Article:

Timely surgical intervention in Masada 4a forearm deformity due to MHE prevents Radial head dislocation and associated morbidity in addition to deformity correction.

Abstract

Introduction: Multiple hereditary exostosis (MHE) is a rare autosomal dominant skeletal disorder widely known as multiple osteochondromatosis that frequently affects the growing ends of long bones. It causes multiple progressive deformities, especially in the wrist, elbow, knee and ankle. The spectrum of features in the forearm includes deformities due to growth arrest of the distal ulnar physis, an increase in the negative ulnar variance, excess radial bowing, an increased radial articular angle, with or without radial head dislocation and restricted pronosupination. Masada's classification applies to forearm deformity in MHE, where Type IVa deformity includes distal osteochondromas of both radius and ulna with ulnar shortening without radial head subluxation or dislocation. It warrants timely surgical intervention to prevent progressive deformity, radial head dislocation and irreversible articular damage.

Aim: The aim of the study was to report the clinical and radiological outcomes of single-stage surgical intervention for Masada Type IVa forearm deformity in children with MHE.

Materials and Methods: Retrospective study of three children with Masada 4 A forearm deformity due to MHE was included in the study. Their mean age was 12.7 years, and they all underwent single stage Ulnar osteochondroma resection, proximal ulnar corticotomy, and gradual ulnar lengthening by distraction osteogenesis using an LRS (Limb reconstruction system) external fixator. All three cases had periodic follow-up with a mean of 27.5 months.

Results: All patients showed high negative ulnar variance needing ulnar distraction lengthening in addition to ulnar osteochondroma excision. Mean ulnar lengthening achieved was 3.6 cm (range: 2.3–4.3 cm). The bowed radius with a mean magnitude of 1.5 in all these cases had a normal range of bow percentage, but with a high radio-capitular articulation inclination angle. High Radio-articular angle of 36° with open physis warranted radial growth modulation in one of the cases, in addition to redo distraction lengthening of the ulna.

Conclusion: Single-stage surgical correction combining distal ulnar osteochondroma excision with Ulnar lengthening by distraction osteogenesis is safe and reproducible for Masada Type IVa deformity in growing children that prevents radial head dislocation. Early intervention confers the greatest benefit of a normal elbow joint in addition to deformity correction that is consistent in radiological and its functional gains.

Keywords: Multiple hereditary exostosis, Masada Type IVa, forearm deformity, ulnar lengthening, distraction osteogenesis, osteochondroma excision, pediatrics orthopedics.

Access this article online

Website: www.jocr.co.in

DOI: https://doi.org/10.13107/jocr.2026.v16.i07.7728

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Submitted: 16/04/2026; Review: 15/05/2026; Accepted: June 2026; Published: July 2026

DOI: https://doi.org/10.13107/jocr.2026.v16.i07.7728

Introduction

Multiple hereditary exostosis (MHE), also referred to as Hereditary multiple osteochondromas, is an autosomal dominant condition characterized by multiple cartilage-capped bony growths over the distal ends of long bones with deformities. It has an estimated population prevalence of 1 in 50,000 to 1 in 100,000 live births [1,2,3,4]. The condition arises from heterozygous germline mutations in either EXT1 (chromosome 8q24.1) or EXT2 (chromosome 11p11-13), both encoding glycosyltransferases essential for heparan sulfate chain elongation [4,5]. Together, these two genes account for pathogenic mutations in roughly 70–85% of clinically diagnosed patients; the remainder either carry variants in other loci or remain genetically unresolved [5]. De novo mutations are not uncommon, occurring in approximately 10–29% of index cases, which has important implications for family counseling and surveillance of apparently unaffected parents and siblings [1].

The forearm is involved in 30–60% of MHE cases, making it the third most frequently affected region after the knee and ankle [6,7]. Ulnar involvement, especially the distal being the primary growth site when tethered by osteochondromas, causes disproportionate shortening of the ulna and hence an increased negative ulnar variance. The mechanical consequence governed by Heuter-Volkman's law causes progressive radial bowing, which affects both the proximal and distal ends. The radius curves in an arc, and to compensate for a fixed distal point, over time distally results in increased Radial articular angle (RAA), with proximal migration of radius causes radio-capitellar joint incongruity and radial head dislocation [2,8]. The rate of deformity progression correlates directly with the duration of the residual skeletal growth, and hence an earlier onset of MHE has a more likely hood of worsening deformities. Its progressive nature can be assessed based on the following parameters. Increased "Ulnar variance" with ulnar shortening of more than 0.8, progressive lateral Radial bowing that exceeds 8.0% of bow magnitude, increased RAA with Radial tilt exceeding the normal range of (15–30°), and subsequent increased carpal slip indicate the degree of carpus uncover. It is not unusual for a child who is manageable at age eight to present with frank radial head dislocation by age eleven or twelve if left untreated [6].

The Masada classification was first described in 1989, the predominant system used in clinical practice for MHE, categorizes forearm deformities into three principal types based on the presence of osteochondromas, their location, the degree of ulnar shortening, and radial head displacement [1,7]. Type I, the most common type (55–61%), involves osteochondroma of the distal ulna with ulnar shortening but without radial head dislocation. Type II involves a distal ulnar osteochondroma with a short ulna and a dislocated radial head. Subtype IIa adds an osteochondroma at the radial head, and Type IIb without a discrete radial head osteochondroma [7,9]. Type III has the primary osteochondroma in the distal radius with relative radial shortening compared to the ulna. Jo's modification of Masada, Type IV includes involvement of both the distal Ulna and Radius without Radial head dislocation in subtype A and with dislocation in subtype B. Type IV A – the pattern seen in all three patients in this series meets at a critical juncture where ulnar shortening is present with increased radial bow and excess radial articular slip angle, but without radial head dislocation. This window is arguably the most surgically essential period, since the radio-capitellar joint retains congruity, prevents radial head dislocation, and the best outcome is achievable through ulnar exostosis excision with gradual distraction osteosynthesis alone [3,8].

Surgical management of MHE forearm deformity has evolved considerably over the past three decades. Early approaches often relied on osteochondroma excision alone, which proved insufficient for meaningful deformity correction [2]. Current

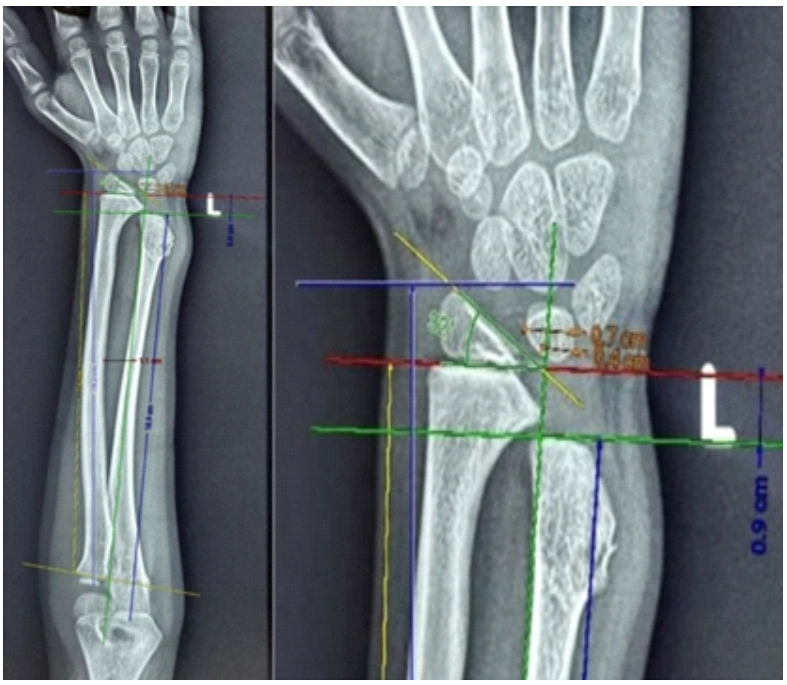

Figure 1: Radiological parameters: (1) Ulnar variance is distance between horizontal lines drawn perpendicular to the linear axis of the forearm at the level of growth plate of radius and ulnar head. (2) Proportionate ulnar length a ratio of ulnar to radial length in cm. (3) Radial articular angle, an angle formed by physeal to epiphyseal lines to the forearm linear axis. (4) Carpal slip a ratio of the uncovered lunate distance on the ulnar side. (5) Radial bow magnitude: The distance at the maximum bow of the radius to the linear axis length.

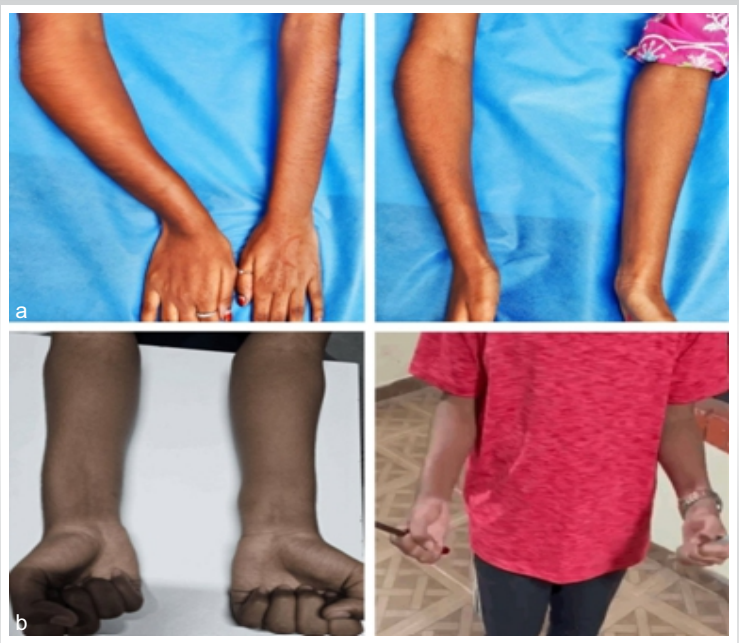

Figure 2: (a-d) Clinical cases with reduced pronation-supination.

strategies from 2019 include ulnar lengthening through distraction osteogenesis, often combined with exostosis resection, as the primary intervention [3,8,10], which prevents radial head dislocation. Corrective radial articular slip angle may be attempted with growth modulation in the younger with open distal radial physis. Radial side of distal radial for corrective osteotomy is reserved for cases with significant fixed radial bowing that persists after ulnar lengthening, provided the physis is closed, and pronosupination is restricted. One-bone forearm fusion is a salvage procedure used only in severe, late-presenting cases or failed prior interventions [2,6]. The optimal timing of surgery, specifically whether to intervene at the point of documented progression or to wait for a threshold deformity angle, remains a matter of clinical debate, though the weight of recent evidence favors intervention before skeletal maturity and before radial head dislocation becomes fixed [6,8,11].

Materials and Methods

This is a retrospective case series conducted at a tertiary care orthopedic center. Three consecutive patients with MHE and progressive forearm deformity meeting the Masada Type IVa criteria were included. Young adults with MHE who had isolated osteochondroma excision and Radial osteotomy were excluded from the study.

Pre-operative evaluation includes general examination, anthropometrics, and assessment of other multiple sites for MHE, including the contralateral side of the forearm. Detailed clinical examination of the forearm side for surgery includes deformity assessment and documentation of elbow, wrist joint range of motion inclusive of forearm pronation-supination (Fig. 1,2). To minimise observer-related measurement bias, all radiological parameters were independently assessed by two authors at different time intervals, and their final values were averaged to minimise the single observer error. Parameters assessed from standard anteroposterior and lateral views radiographs include Ulnar variance (in mm), proportional ulnar length (<0.9 a strong indicator of progressive deformity), maximum radial bowing (Radial bow magnitude in cm), radial bow percentage (percentage of radial bow magnitude to radial length), radio-articular angle (RAA in degrees°) [9] (Fig. 1). Functional outcome assessment was performed using QuickDASH scoring during their latest clinical follow-up evaluation.

All three patients underwent a single-stage procedure under general anesthesia: Procedure includes partial resection of the ulnar osteochondroma followed by proximal ulnar corticotomy at the proximal one-third of the diaphysis, and application of a rail (LRS) external fixator for distraction osteogenesis [3,10]. Distraction commenced after a standard 5-day latency period at a rate of 0.25 mm 4 times daily (1 mm/day total), and was continued until adequate ulnar length was restored based on the high negative Ulnar variance and proportionate ulnar length. Close clinical and radiographic reviews were carried out during the distraction and consolidation phases. All patients underwent supervised distraction protocol and dedicated post-operative hand physiotherapy for guided rehabilitation, focusing on forearm rotation and functional recovery. Mean follow-up was 27.3 months (range: 24–28 months).

Results

Radiological parameters for all three cases were summarized in Table 1 for the operated side. All these cases showed a high negative ulnar variance with a mean of 2.26, but 1.5 for non-operated side. Mean ulnar length gain achieved postoperatively was 3.6 cm (range: 2.3–4.3 cm). The Radial bow percentage remained within the threshold of 9% both in pre and post-operative follow-up assessment. The mean pre-operative RAA was 30.6, and in the post-operative measured a mean of 36°. The contralateral forearm showed negative ulnar variance, but radial bow and RAA were within normal threshold. Two of the three patients reached skeletal maturity during follow-up with maintained radiological correction and functional improvement (Fig. 3). The youngest patient, with significant residual growth potential at 24 months follow-up, developed progressive radial articular angulation deformity. She underwent a second distraction osteogenesis with distal radial growth modulation at the age of 11 years.

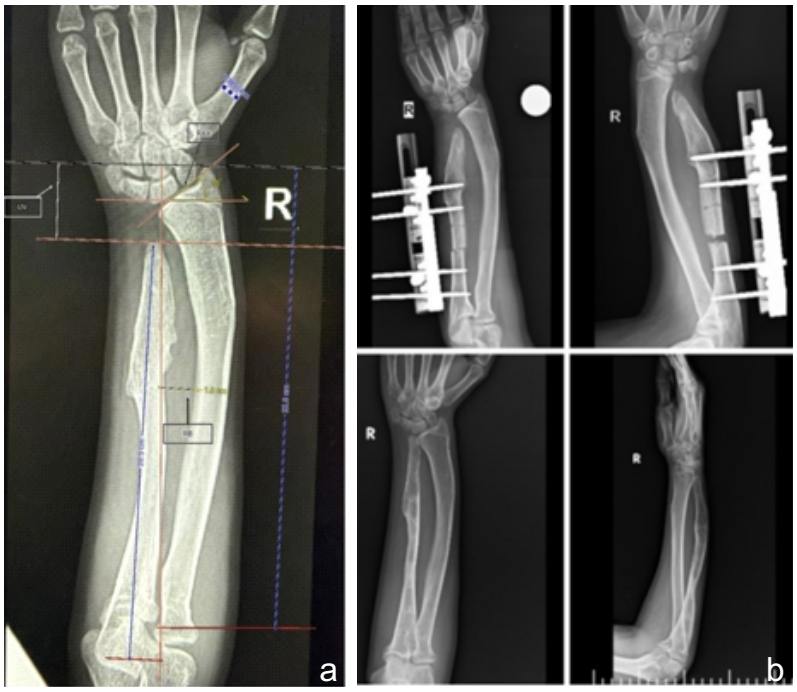

Figure 3: (a) Pre-operative images of 14 years old with distal ulnar osteochondroma high ulnar variance = 2.3, radial bow magnitude of 1.8 cm, bow percentage = 7.8. (b) Post-operative radiographs following osteochondroma excision with ulnar corticotomy; distraction phase in progress. Radiographs at 18-month follow-up.

Literatures lack disease- specific outcome scores for MHE. Functional assessment using the QuickDASH score at final follow-up demonstrated satisfactory upper limb function in all patients. Final follow-up QuickDASH scores were 14, 16, and 20, respectively. Objective assessment in the form of forearm rotations of the elbow was assessed (Table 1). The grip strength was challenging in children and was not attempted.

No major literature reported complications such as pin tract infection, delayed union, regenerate deformity, neurovascular compromise, or loss of correction were encountered during follow-up in our series.

Additional procedures

The youngest patient in the series, at the age of 9 years, underwent unilateral deformity correction with excision of the ulnar osteochondroma and ulnar lengthening. At 24 months follow-up, with residual growth left despite a negative ulnar variance of 0.7 had a high RAA of 36°. This was considered a risk of radial head dislocation with residual growth and potential for the radial bow progression. Hence, she was subjected to radial growth modulation and redo ulnar corticotomy and lengthening to compensate for the residual radial growth (Fig 4).

Discussion

Forearm deformity in MHE carries a progressive nature with skeletal growth. The relative ulnar shortening with progressively increased radial bow contributes to radial head dislocation in type 4a. The added peak growth velocity has the potential risk of radial head dislocation and permanent disability in addition to a cosmetically visible deformity [6,11]. EXT1 and EXT2 mutations clinically differ due to the genotype-severity relationship. Patients harboring EXT1 variants tend to develop

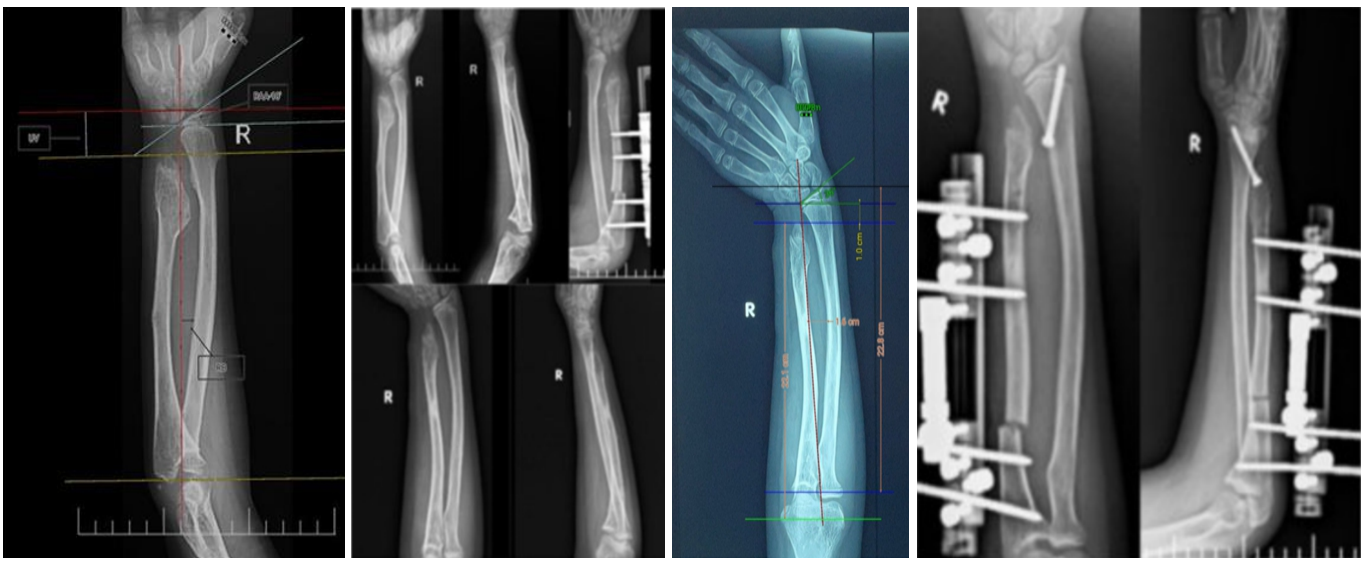

Figure 4: (a) Pre-operative anteroposterior, lateral radiograph Masada Type IVa deformity pattern in a 9-year-old. Increased Ulnar variance of 2.2 and radial bow percentage of 5.9, but with radial articular angle (RAA) = 18°. (b) Post-operative anteroposterior and lateral radiographs showing ulnar corticotomy with distraction osteogenesis and gradual ulnar lengthening. Eighteen-month follow-up radiographs. (c) RAA –36° at 24 month. (d) 2nd stage corticotomy – Redo distraction osteogenesis with growth modulation for the distal radius.

Table 1: Radiological and functional parameters – pre-operative and post-operative comparison

Parameter	C1 Pre	C1 Post	C2 Pre	C2 Post	C3 Pre	C3 Post
Radial length (cm)	22.8	23.1 cm	22.7	25.5	20.1 cm	22.8
Ulnar length (cm)	20.5	22.8	20.4	24.7	17.9 cm	22.1
Ulnar variance (cm)	2.3	0.8	2.3	0.8	2.2	0.7
Proportional ulnar length	1.11	0.98	0.89	0.96	0.89	0.96
Radial bowing (cm)	1.8	2.1	1.5	1.4 cm	1.2 cm	1.6
Radial bow percentage	7.8	9	6.6	5.4	5.9	7
Supination	0–80°	0–80°	0–40°	0–70°	0–60°	0–70°
Pronation	0–20°	0–60°	0–30°	0–70°	0–50°	0–70°
Radio-articular angle (°)	38°	32°	36°	40°	18°	36°
Quick Dash score	-	14	-	16	-	20

more numerous and more deforming exostoses, with greater carpal slip angles and higher rates of forearm involvement [1,4]. This distinction is not merely academic but helps in robust surveillance and planning early surgical intervention as per the systematic review by Pedrini et al. Previous studies have shown that untreated progressive ulnar shortening in MHE may lead to worsening radial bowing, restriction of forearm rotation, and eventual radial head dislocation, supporting the role of early surgical intervention in Masada Type IVa deformity [1,6,7].

Cases in this series were Masada 4a types where the radio-capitellar joint is prone to dislocation, reflecting the clinical value of active surveillance in known MHE patients during the growth years.

The single-stage procedure of combining the ulnar osteochondroma excision with corticotomy and immediate distractor application for lengthening will be the single-stage approach and is associated with superior alignment, in addition to prevention of radial head dislocation [3,10].

The osteotomy level chosen in all three cases was the proximal one-third of the ulnar diaphysis, which is the only possible zone consistently shown to have the most reliable regenerate bone formation in distraction series, a finding reproduced in recent Ilizarov-based ulnar lengthening studies [10,12]. The recommended metaphyseal site for corticotomy is unsuitable in children, with available proximal space being inadequate for distractor pin placements. Distraction at 1 mm/day (0.25 mm 4 times daily) following a 5-day latency phase aligns with Ilizarov's original tension-stress principles and remains the most widely adopted protocol in pediatrics forearm lengthening [10].

The question of corrective radial osteotomy in addition to ulnar lengthening is often decided based upon the residual growth potential and the bow magnitude. In Type IVa deformity, where radial bowing is present but the radiocapitellar joint retains congruity, ulnar lengthening alone generally achieve radiocapitellar reduction without addressing the radius directly [2,8]. To which our results corroborate without radial osteotomy. Persistent radial head prominence after adequate ulnar distraction will be an indication for radial corrective osteotomy on the same sitting if proven on the fluoroscopy screening before removal of the distractor [2]. Timely surgical intervention helped preserve radio capitellar congruity and forearm biomechanics by preventing progressive radial head subluxation/dislocation.

Age at intervention emerged as a functionally relevant variable within our small cohort. The 9-year-old (Case 3) achieved the most complete functional recovery at 18 months, with the broadest pronation, supination arc, and the greatest subjective improvement in daily activities. Both older patients (aged 14 and 15 years) also benefited meaningfully, though residual pronation restriction is an anticipated sequela in adolescents approaching skeletal maturity, as noted in comparable case series [2,5]. Two of the three patients reached skeletal maturity during follow-up with maintained radiological correction and functional improvement. The youngest patient, with significant residual growth potential, developed progressive radial articular deformity and subsequently underwent repeat distraction osteogenesis with distal radial growth modulation. This highlights the importance of continued surveillance in younger children with substantial residual growth. The satisfactory QuickDASH scores at final follow-up correlated with improved forearm rotation and functional upper limb use following deformity correction.

Complications inherent to external fixator use include pin tract

infection, premature consolidation, and regenerate fracture, which were not encountered in this series, though the cohort is clearly too small to characterize the complication rate with any confidence. Published rates of pin tract infection in pediatric forearm distraction range from 15% to 40%, most of which are superficial and responsive to oral antibiotics without frame removal [12,13].

From a quality-of-life perspective, MHE carries a measurable burden beyond the deformity itself. The potential for radial head dislocation, activity restriction, school absence, and psychological impact was documented across patient-reported outcome studies [13]. Restoration of forearm rotation, in particular, has direct implications for activities of daily living, including feeding, grooming, and writing [6]. Deformity recurrence during subsequent growth spurts needs recognition and necessitates surveillance through to skeletal maturity [11].

Limitations of the study

This study is limited by its retrospective design and small sample size of only three Masada Type IV A cases, and hence, statistical significance could not be drawn. Being a single-center study, the outcomes may reflect institutional expertise and potential selection bias. Genetic analysis was not performed in these patients, and henceforth the genetics could not be correlated with their morphometrics in MHE. The short-term follow-up limits the assessment of long-term outcomes. However, early surgical intervention successfully prevented radial head dislocation and improved forearm pronation–supination, only long-term follow-up can show the effects of altered elbow biomechanics.

Conclusion

Single-stage surgical correction is a safe and reproducible technique for Masada Type IVa forearm deformity in children with MHE. Across this series, all three patients achieved meaningful radiological realignment and prevention of radial head dislocation with functional improvement at short-to-medium-term follow-up. Correction at a younger age demonstrated the need for longer-term follow-up through skeletal maturity to prevent late-onset bow deformity and its consequences.

Clinical Message

Early surgical intervention for Masada Type IVa forearm deformities in Multiple Hereditary Exostosis (MHE) can prevent progressive radial head dislocation and associated functional morbidity while effectively correcting the deformity. Once the deformity crosses the acceptable threshold, surgical treatment becomes necessary to restore and maintain normal elbow biomechanics by preventing or addressing radial head dislocation.

Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given the consent for his/ her images and other clinical information to be reported in the journal. The patient understands that his/ her names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Conflict of interest: Nil **Source of support:** None

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14. Genotype-phenotype correlation study in 529 patients with multiple hereditary exostoses: identification of "protective" and "risk" factors

Elena Pedrini 1, Ivy Jennes, Morena Tremosini, Annamaria Milanesi, Marina Mordenti, Alessandro Parra, Federica Sgariglia, Monia Zuntini, Laura Campanacci, Nicola Fabbri, Elettra Pignotti, Wim Wuyts, Luca Sangiorgi

Conflict of Interest: Nil
Source of Support: Nil

Consent: The authors confirm that informed consent was obtained from the patient for publication of this article

How to Cite this Article

Sangai N, Sitsabesan C, Menon G, Ponraj DGS. Outcomes of Surgical Treatment for Masada Type lVa Forearm Deformity in Children with Multiple Hereditary Exostosis: A Case Series. Journal of Orthopaedic Case Reports 2026 July;16(07):484-490.