Midterm Outcomes of Isolated Medial Patellofemoral Ligament Reconstruction for Patellar Instability in Ehlers-Danlos Syndrome

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Background: Patellar instability is frequently encountered in patients with Ehlers-Danlos syndrome (EDS). The clinical outcomes of isolated medial patellofemoral ligament reconstruction (MPFLR) for patellar instability in patients with EDS are unknown.

Purpose: To evaluate midterm clinical outcomes of isolated MPFLR for patellar instability in patients with EDS and factors affecting these outcomes.

Study Design: Case series; Level of evidence, 4.

Methods: In a retrospective study, 31 patients (n = 47 knees) with EDS and patellar instability who underwent isolated MPFLR for recurrent patellar instability between 2008 and 2017 and had a minimum 2-year follow-up were identified. Preoperative radiographic images were measured for anatomic risk factors. Clinical outcomes—including postoperative complications—were evaluated. Factors associated with MPFLR failure were identified. Postoperative patient-reported outcomes (PROs)—including the pediatric version of the International Knee Documentation Committee, the Kujala score, the Hospital for Special Surgery Pediatric Functional Activity Brief Scale, the Banff Patellofemoral Instability Instrument 2.0, and the Knee injury and Osteoarthritis Outcome Score—were collected, and factors affecting PRO scores were analyzed.

Results: The mean age of the cohort was 14.9 $\pm$ 2 years. At a mean follow-up of 7.2 years, 18 of 47 (38.3%) knees required reoperations, of which 9 of 47 (19.1%) knees required revision stabilization for recurrent patellar instability. Also, 7 of 31 knees (22.6%) with autografts failed compared with 2 of 16 (12.5%) with allografts (P = .69). For autografts, 6 of 17 (35.3%) failures occurred with gracilis, but 0 of 13 (0%) occurred with semitendinosus (P = .02). Compared with patients without failures, patients with failed primary MPFLR were significantly younger (P = .0005) and were able to touch the palm to the floor with their knees extended (P = .03). For radiographic parameters, the patellar height and tilt were significantly higher in the failure group. The postoperative PROs were suboptimal at a mean follow-up of 5.2 years. All but 1 patient were satisfied with the final outcome.

Conclusion: At the midterm follow-up, 38.3% of patients with EDS required further surgery after isolated MPFLR for patellar instability; half of these revisions (19.1%) were to address recurrent instability. Recurrent instability after isolated MPFLR was more likely in younger patients and those who could touch the palm to the floor with their knees extended. Postoperative PROs were inferior; nonetheless, patient satisfaction was high.

Keywords: Ehlers-Danlos syndrome; knee; medial patellofemoral ligament reconstruction; patella; pediatric sports medicine

Ehlers-Danlos syndrome (EDS) is a heterogeneous group of connective tissue disorders characterized by joint hypermobility, skin hyperextensibility, and tissue fragility.[20] The prevalence of EDS is estimated to be 1 in 5000 individuals worldwide.[40] The features of EDS are secondary to abnormal collagen due to mutations in several different genes. A Beighton score of $\geq$5 out of a possible 9 points is used to define generalized joint hypermobility and is one of the major criteria for EDS diagnosis.[14,20] The other criteria are as follows: systemic manifestation of generalized

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connective tissue disorder; positive family history, musculoskeletal pain/joint instability; and exclusion of other connective tissue disorders, such as rheumatologic conditions or skeletal dysplasia.[20]

Patellar instability is common in children and adolescents with EDS. In a retrospective review of 205 patients with EDS between the ages of 6 and 19 years old, the most common site of complaint was the knee, primarily involving the patella.[36] In a survey, 57% of patients with EDS reported patellar dislocation, and 93% reported generalized joint pain.[1] In another survey, 81.5% of patients with EDS reported knee pain, and 40.7% reported patellar instability.[27] Sheehan et al[35] found significant alterations in patellofemoral kinematics in the EDS population. The management of patellar instability in patients with EDS is challenging because of inherent tissue laxity, and it is further compounded by the presence of chronic pain, generalized hyperalgesia, fatigue, muscle weakness, muscle imbalance, and psychological issues.[7,28,30,38,42]

Medial patellofemoral ligament reconstruction (MPFLR) is an acceptable treatment option to address patellar instability in the young population. In a meta-analysis, the risk of recurrent instability after an isolated MPFLR was 1.2%, and the reoperation risk was 3.1% in the normal population.[31] Similar encouraging results of isolated MPFLR have been reported in skeletally immature patients.[33] However, to our knowledge, clinical outcomes of isolated MPFLR for patellar instability in the EDS population have not yet been reported.

The purpose of the present study was to evaluate the demographic and clinical characteristics of patellar dislocation in patients with EDS and report on the midterm outcomes of isolated MPFLR for patellar stabilization in this population.

METHODS

Our institutional review board approved the protocol for this study. As part of a retrospective study, a hospital surgical database was queried to identify patients who underwent knee extra-articular ligament reconstruction (Current Procedural Terminology code 27427) between 2008 and 2017. A total of 532 knees were identified, of which 510 had MPFLR procedures. The electronic medical records were reviewed to identify patients with a concomitant EDS diagnosis. Most patients were referred from the hypermobility/EDS clinic—a specialty clinic within the Division of Human Genetics at our institution. A total of 56 knees (n = 37 patients) with EDS were identified, representing 9.4% of all patients who underwent MPFLR during this period. Of these, 50 knees (n = 34 patients) had a minimum 2-year clinical follow-up. Two patients (n = 2 knees) with MPFLR and concurrent tibial tubercle osteotomy and 1 patient with vascular EDS were excluded. This formed the final cohort of 47 knees (n = 31 patients) with EDS who underwent isolated MPFLR for patellar instability with a minimum 2-year follow-up. IRB waived documented consent requirement however verbal consent was collected when patients were contacted about the one time follow up survey.

The electronic medical records of these patients were reviewed to determine the demographic factors (age, sex, laterality, and Beighton score) and any previous treatment. Preoperative radiographs and magnetic resonance imaging (MRI) were evaluated and measured for 4 anatomic risk factors, including trochlear dysplasia, patellar height, patellar tilt, and tibial tubercle–trochlear groove (TT-TG) distance using previously described methods.[3,9] Trochlear dysplasia was classified as none, low grade (Dejour type A), or high grade (Dejour types B, C, or D).[17] The patellar height was evaluated on lateral radiographs using the Caton-Deschamp index (CDI), with values >1.2 indicating patella alta. The patellar tilt was evaluated on an axial MRI slice, with values >20° considered abnormal. The TT-TG distance was calculated using axial MRI slices, with values >20 mm considered abnormal. MRIs were evaluated for assessment of physeal status. The patient was considered skeletally mature if any part of the tibial or femoral physis was closed.[8]

The indication for surgery was recurrent patellar instability in all patients. The operative records were reviewed for MPFL graft type and other concomitant procedures. Isolated MPFLR was defined as MPFLR with free tendon graft without any concomitant bony procedures. Patients with lateral retinacular release and/or chondroplasty were included. MPFLR was performed using a standard technique that has been previously described.[23] A single 3.5-mm patellar tunnel was created in the superior half of the patella, and the tendon graft was looped through the tunnel and over a bone bridge. Both ends of the graft were whipstitched with a nonabsorbable suture. The MPFL femoral attachment point was identified using fluoroscopy at the Schöttle point.[32] This point was moved distal to the femoral physis in skeletally immature patients. A 5.5 to 6 mm femoral tunnel was drilled as per the size of the doubled graft. The graft was passed between layers

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Ethical approval for this study was obtained from Cincinnati Children's Hospital Medical Center (ref No. 2017-3058).

TABLE 1
Previous Surgeries and Associated Procedures[a]

Previous Surgeries	Associated Procedures During MPFLR
Lateral retinacular release, n = 3	Patellar chondroplasty, n = 13
VMO reefing, n = 2	LFC chondroplasty/pick arthroplasty, n = 5
Arthroscopic removal of loose bodies, n = 2	Patellar osteochondral fracture fixation, n = 1
MPFL repair, n = 1	Removal of hardware from previous surgery, n = 1
Tibial tubercle osteotomy, n = 1	Lateral retinacular lengthening, n = 1
Synovectomy, n = 1	Lateral meniscus debridement, n = 1
Total: 10 surgeries in 8 knees	Total: 22 surgeries in 20 knees

[a]LFC, lateral femoral condyle; MPFL, medial patellofemoral ligament; MPFLR, medial patellofemoral ligament reconstruction; VMO, vastus medialis obliquus.

2 and 3 on the medial side of the knee and then into the femoral tunnel. Femoral fixation was performed using an interference screw with the knee in 45° to 60° of flexion. Standard postoperative physical therapy protocol was utilized for all patients—including weightbearing as tolerated with crutches and a knee immobilizer until quadriceps activation was achieved. Return to full activities was allowed after 6 months once functional testing was performed.

Any intra- or postoperative complications were noted. Failure was defined as the need for any subsequent intervention. Patients with recurrent patellar instability after isolated MPFLR were further analyzed based on age, laterality, type of graft, Beighton score, and radiographic parameters. Intraoperative fluoroscopy images were reviewed to evaluate the femoral tunnel position.

At the final follow-up, patients were asked to complete the patient-reported outcomes (PROs)—including the pediatric version of the International Knee Documentation Committee (Pedi-IKDC), the Kujala score, the Hospital for Special Surgery Pediatric Functional Activity Brief Scale (HSS Pedi-FABS), the Banff Patellofemoral Instability Instrument 2.0 (BPII), and the Knee injury and Osteoarthritis Outcome Score (KOOS). All of these PROs have been validated in pediatric and adolescent patients.[37] The best possible score for the HSS Pedi-FABS was 30, and the best possible score for all other PROs was 100. PROs were collected using the REDCap electronic data capture system (REDCap), hosted at our institution. PRO scores were analyzed based on age, laterality, graft type, and Beighton score. Satisfaction with overall treatment and final outcome was also assessed at the latest follow-up.

Statistical Analysis

Continuous variables were reported as means with standard deviations, and comparisons between groups for continuous variables were performed using the Student *t* test. Categorical variables were described as frequencies with percentages, and comparisons between groups for categorical variables were performed using the Fisher exact test. $P < .05$ was considered significant. Statistical analysis was done using Microsoft Excel (Microsoft; Version 2402).

RESULTS

The mean age of the cohort was 14.9 ± 2 years (range, 11-19 years). Of the 31 patients included, 27 (87.1%) were female. Eleven (23.4%) knees were classified as skeletally immature. The left knee was involved in 9 patients, the right knee was involved in 6 patients, and 16 patients (51.6%) had bilateral knee involvement. Also, 8 of 47 (17%) knees had previous surgeries, summarized in Table 1. The mean Beighton score for the cohort was 6.8 ± 1.5. There were 26 patients with a Beighton score of 9 of 9. For the Beighton score evaluation related to lower extremities, 32 knees had genu recurvatum >15°, and 26 patients could place their palms on the floor with their knees extended.

Preoperative radiographs and MRI showed high-grade trochlear dysplasia in 20 of 47 (42.6%) knees. The mean patellar height was 1.3 ± 1.2, with 23 of 47 (48.9%) having a CDI of >1.2. The mean patellar tilt was 18.9° ± 7.2°, with 22 of 47 (46.8%) having a tilt of >20°. The mean TT-TG distance was 13.2 ± 5.4 mm, with 6 of 47 (12.8%) having a distance of >20 mm.

MPFLR was performed using a hamstring allograft (n = 16 knees) or an autograft (n = 31 knees: 17 gracilis, 13 semitendinosus, and 1 quadriceps tendon). The graft choice was based on shared decision-making between the surgeon and the patient/family. Twenty knees had concomitant procedures performed at the time of MPFLR (Table 1). For the 16 patients with bilateral knee involvement, 3 patients had both knee operations at the same time, and 13 patients had staged surgeries. There were no intraoperative complications. The mean follow-up for the entire cohort was 7.2 ± 3.4 years (range, 2-13 years).

Postoperative complications and unplanned subsequent surgeries are listed in Tables 2 and 3. The overall number of knees that required further surgery was 18 of 47 (38.3%), of which 9 of 47 (19.1%) knees had recurrent instability after MPFLR and required revision stabilization surgery (Table 2). Except for 1 knee, all failures were related to an atraumatic mechanism of injury. Four knees had occasional instability episodes; however, these could not be objectively verified, and patients did not seek further treatment.

TABLE 2
Revision Surgeries for Recurrent Instability[a]

Patient Knee	Age, y. mo	Sex	U/B	Graft	BS	Palm	GR	No. of Revision Surgeries (Time Since Index Surgery, y)
1	10.2	F	B	Allo	6	Yes	No	1: TTO and LRL (2)
2	10.10	F	B	Allo	6	Yes	Yes	1: TTO and LRL (3)
3	11.3	F	B	Gr	9	Yes	Yes	1: Trochleoplasty and MPFLR using allograft (2)
4	11.7	F	B	Gr	9	Yes	Yes	1: Trochleoplasty and MPFLR using allograft (2)
5	15.7	F	U	Q	6	No	Yes	2: MPFLR using allograft and patellar chondroplasty (1); trochleoplasty and MPFLR using autograft (5)
6	13.7	M	B	Gr	7	Yes	No	3: OATS to LFC (1); MPFLR using allograft (2); patellar chondroplasty (3)
7	14.4	M	B	Gr	7	Yes	No	2: MPFLR using allograft (1); patellar chondroplasty (2)
8	12.9	F	U	Gr	6	Yes	Yes	1: MPFLR using allograft (1)
9	15.2	M	B	Gr	7	Yes	No	1: MPFLR using allograft (2)

[a]Allo, allograft; B, bilateral; BS, Beighton score; F, female; GR, genu recurvatum of involved knee; Gr, gracilis autograft; LFC, lateral femoral condyle; LRL, lateral retinacular lengthening; M, male; MPFLR, medial patellofemoral ligament reconstruction; OATS, osteoarticular transfer system; Palm, the ability to place the palm on the floor with knees extended; Q, quadriceps tendon autograft; TTO, tibial tubercle osteotomy; U, unilateral.

TABLE 3
Complications and Unplanned Surgeries After Isolated MPFLR, Excluding Revision Surgeries for Recurrent Instability[a]

Patient Knee	Age, y. mo	Sex	U/B	Graft	BS	Complications	No. of Surgeries (Time Since Index Surgery, y)
1	19	F	U	ST	9	DVT, wound dehiscence, arthrofibrosis	2: Wound closure (6 wk); MUA (2 mo)
2	13.10	F	B	Gr	7	Pain	1: Patellar chondroplasty (9 y)
3	13.10	F	B	Gr	7	Pain	2: Patellar chondroplasty (9 y); patellar chondroplasty (10 y)
4	14.8	F	B	ST	5	Pain	1: Saphenous neuroma excision (10 y)
5	17.8	F	U	Allo	4	Pain	1: Femur interference screw removal (9 mo)
6	16.6	F	B	Gr	7	Pain	2: Patellar chondroplasty (5 y); patellar chondroplasty (8 y)
7	18.6	F	B	Gr	7	Pain	1: Patellar chondroplasty (6 y)
8	16.11	F	B	Allo	8	Deformity	1: Reversal of TTO (6 y)
9	15.6	F	B	Gr	5	Pain	1: Patellar chondroplasty (2 y)

[a]Allo, allograft; B, bilateral; BS, Beighton score; DVT, deep venous thrombosis; F, female; Gr, gracilis autograft; MPLFR, medial patellofemoral ligament reconstruction; MUA, manipulation under anesthesia; ST, semitendinosus autograft; TTO, tibial tubercle osteotomy; U, unilateral.

Patients with recurrent instability after isolated MPFLR who required revision stabilization surgery were further analyzed (Table 4). These patients were younger and were able to touch the palm to the floor with their knees extended ($P < .05$). There was a trend toward a higher failure rate in those with bilateral involvement but it was not statistically significant ($p = 0.65$). Compared with allografts, MPFLR with an autograft had a higher failure rate though it was not statistically significant ($p = 0.69$). In the autograft failure group, 6 of 17 (35.3%) knees with gracilis autografts failed, but none of the 13 knees with semitendinosus autografts failed; this was statistically signiciant ($p = 0.02$). For preoperative radiographic parameters, the patellar height and tilt were significantly higher in the failure group. Evaluation of intraoperative fluoroscopy images showed accurate positioning of the femoral tunnel at the Schöttle point in all skeletally mature patients. The femoral attachment point was moved distal to the distal femoral physis for skeletally immature patients. None of the skeletally immature patients had any growth disturbances at the latest follow-up.

Regarding PROs, 38 of 47 (80.9%) responded at a mean follow-up of 5.2 ± 1.9 years postoperatively. The mean Pedi-IKDC score was 69.2 ± 19.8, the Kujala score was 75 ± 20.1, the HSS Pedi-FABS was 5.9 ± 6.7, the BPII 2.0 score was 58.8 ± 25.1, the KOOS-Pain score was 76.1 ± 22.5, the KOOS-Symptoms score was 71.2 ± 19.4, the KOOS Activities of Daily Living score was 82.3 ± 21, the KOOS-Sport score was 64.7 ± 29.4, and the KOOS Quality of Life score was 56.5 ± 25 (Appendix Table A2). There were no statistically significant differences in PROs when compared between subgroups related to age (≤15 or >15 years), knee involvement (unilateral or bilateral), Beighton score (≤7 or >7), or graft type

TABLE 4
Comparison Between Knees With and Without Revision Stabilization Surgery[a]

	No revision (n = 38 knees)	Revision (n = 9 knees)	% failed	*p*
Age (y)	15.4 ± 1.8	12.9 ± 1.8	NA	**p = 0.0005***
Female (n = 27 patients)	21	6	22.2%	p = .06
Male (n = 4 patients)	1	3	75%	
Unilateral (n=15 patients)	13	2	13.3%	p = 0.65
Bilateral (n=16 patients)	12	4 (7 knees)	25%	
Beighton Score	6.7 ± 1.5	7 ± 1.5	NA	p = 0.59
L Recurvatum (n=32 knees)	27	5	15.6%	p = 1
R Recurvatum (n=32 knees)	27	5	15.6%	
Palm to Floor (n=26 patients)	18	8	30.8%	**p = 0.03***
Allograft (n=16 knees)	14	2	12.5%	p = 0.69
Autograft (n=31 knees)	24	7	22.6%	
Gracilis autograft (n = 17 knees)	11	6	35.3%	**p = .02***
Semitendinosus autograft (n = 13 knees)	13	0	0	
Radiographic Parameters (Knees)				
High-grade TD	16 (42.1%)	4 (44.4%)	NA	p = 0.9
CDI	1.2 ± 0.2	1.5 ± 0.2	NA	**p = 0.0002***
Patellar tilt	17.3 ± 6.5	25.2 ± 6.9	NA	**p = 0.002***
TT-TG distance	12.9 ± 5.7	14.5 ± 3.8	NA	p = 0.43

[a]Data are reported as mean ± SD, n (%), or No. of knees unless otherwise indicated. Bold *P* values indicate statistically significant differences between revision and no revision groups ($P < .05$). CDI, Caton-Deschamps Index; L, left; R, right; NA, not applicable; TD, trochlear dysplasia; TT-TG, tibial tubercle–trochlear groove.

(allograft or autograft, gracilis autograft or semitendinosus autograft) (Appendix Table A1). All but 1 patient were satisfied with the final outcome, irrespective of the need for subsequent surgeries.

DISCUSSION

The most important finding of our study is that isolated MPFLR can be effective for patellar stabilization in patients with EDS with a 19.1% recurrent instability rate at a mean 7-year follow-up. Factors associated with failure were younger age and ability to place the palm on the floor with knees extended. There was a trend for higher failure rates in those with bilateral involvement and with the use of the gracilis autograft. Another 19.1% required subsequent surgeries, the most common being patellar chondroplasty. Thus, the total reoperation rate was 38.3%. Postoperative PROs were suboptimal, but patient satisfaction was high.

Hiemstra et al[11] evaluated the effect of generalized joint hyperlaxity (GJH) on outcomes after isolated MPFLR for patellar instability. They found that of 92 patients with a positive (≥4) Beighton score, 3 patients had recurrent instability, compared with 4 patients with recurrent instability in 75 patients with a negative (<4) Beighton score. There was no evidence of a relationship between GJH and disease-specific quality of life scores or objective functional outcomes, whereas, in our study, there was a significantly higher rate of recurrent instability and inferior PROs in patients with EDS. The following variances in study methodology can explain the difference in outcomes. First, our cohort represented a consecutive series of patients with EDS who underwent isolated MPFLR irrespective of underlying anatomic risk factors, whereas Hiemstra et al[11] excluded 59 patients from their cohort, as they had other procedures besides MPFLR for correction of anatomic risk factors. Second, the mean age of our cohort was much younger at 14.9 years compared with 24.1 years, and it is well known that younger age is a major risk factor for patellar instability.[13,16] Third, Hiemstra et al[11] did not use a knee-specific outcome instrument (eg, Kujala or IKDC) for evaluation; thus, it is likely that their patients may have knee-related limitations, but they may not have affected their overall quality of life. Fourth and perhaps most important, patients with EDS comprise a unique and more complex subset of patients compared to patients with GJH.

In another study, Howells and Eldridge[12] compared the outcomes of isolated MPFLR in 25 patients with hypermobility (Beighton score, ≥6) with those of a control group of 50 patients without hypermobility (Beighton score, <4). There were no recurrent dislocations in either group. The functional outcome scores were improved in patients with hypermobility but to a lesser extent than in controls. However, the follow-up in their study was significantly limited, with a minimum follow-up of 6 months and a mean follow-up of 16 months. The higher rate of recurrent instability in our study could be due to much younger mean age (14.9 vs 25.4 years), longer mean follow-up in our study (5.2 years vs 16 months), longer minimum follow-up in our study (2 years vs 6 months), and our distinct group of patients with EDS. In a 2022 study, Reddy et al[25] reported a 9% recurrent instability rate with the use of allografts for isolated MPFLR in children and adolescents with hypermobility.

The present study sheds light on the graft options for MPFLR in patients with EDS. Although not statistically significant, the 22.6% failure rate with autografts (primarily gracilis) was almost double compared with the 12.5% failure rate with allografts in our cohort. Although a meta-analysis showed no significant differences between revision rates for autograft and allograft,[44] Kumar et al[15] reported 6 of 23 (26.1%) failures in the autograft group compared with 3 of 36 (8.3%) failures in the allograft group in 59 adolescent patients at a mean follow-up of 4.1 years after MPFLR. The authors noted worsening outcomes with longer follow-ups in the autograft group but not in the allograft group, possibly because of graft attrition over time. Similarly, Hendawi et al[10] reported on 56 pediatric patients with MPFLR and noted a significantly higher failure rate (28.6%) in the autograft group compared with no failures in the allograft group at a mean of 13.8 months postoperatively. The authors postulated smaller graft size and increased laxity in younger patients as possible reasons for increased failures of autografts in their series. In a systematic review, the range of failure rate of autografts (0%-28.6%) was worse than that of allografts (0%-8.3%).[2] The authors concluded that allografts should be favored over autografts because of lower failure rate, shorter operative time, and decreased donor-site morbidity. In EDS, tendon elongation is much greater, and tendon stiffness is much lower than in controls.[28] Clinically, increased hamstring elasticity and stretch are manifested by the ability to put the palm on the floor with knees extended. In the present study, there was a significantly higher failure rate in patients with EDS who were able to put their palms on the floor with their knees extended. In such circumstances where hamstring tendons could stretch or elongate significantly, strong consideration should be given to the use of an allograft. If an autograft is chosen for MPFLR, then consideration should be given to use the larger semitendinosus tendon instead of the smaller gracilis tendon as significant higher failure rate was noted for gracilis autograft in the current study.[2]

Patients of younger age and with bilateral instability had higher failure rates of isolated MPFLR in the present study. This finding is not surprising, as both are known risk factors for recurrent patellar instability in the non-EDS population.[13,23] Tissue elasticity and joint hyperlaxity are more prevalent at a young age and progressively decrease with increasing age. There is a change in collagen type with a higher proportion of collagen III compared with collagen I with increasing age.[24] Because of increased joint laxity at a young age, the cutoff Beighton score for diagnosis of GJH in children is 6, decreasing to 5 for adolescents.[14] Besides young age, bilateral instability may suggest associated underlying patellofemoral dysplasia.

There was a high (42.6%) prevalence of high-grade trochlear dysplasia in our cohort, but it was not statistically different in the failure group. Trochlear dysplasia may result from insufficient loading patterns in the patellofemoral joint secondary to hyperlaxity.[39] There was a statistically significant increase in patellar height and tilt in patients who had failed isolated MPFLR in our cohort. A cautious approach is warranted when correction of the patellar tilt is considered in patients with hyperlaxity, as lateral retinacular release can lead to iatrogenic medial instability. The approach of isolated MPFLR for patellar instability is supported by several studies that have reported satisfactory results after isolated MPFLR in the presence of trochlear dysplasia, patella alta, or increased TT-TG distance.[4,6,18,31,41]

The family should be counseled about the higher rates of failure of isolated MPFLR in this subset of patients with EDS. Once skeletally mature, bony procedures for correcting underlying dysplasia (eg, tibial tubercle osteotomy or trochleoplasty) could be considered, as were performed in 5 of 9 revision surgeries in our cohort. Such procedures are contraindicated in younger, skeletally immature patients.

Many of our patients had ongoing pain issues related to their knees. This finding is similar to a study of 44 patients with EDS, which reported chronic knee pain in 85% of patients.[43] In another study, 93% of patients with hypermobile EDS had joint pain or arthralgia, and 40% of patients required some aid for mobility.[1] Pain is usually multifactorial and may be influenced by psychological issues, increased fatigue, muscle imbalance, and muscle weakness. Patients with EDS can also have associated fibromyalgia.[26] Besides pain, other potential postoperative complications in the literature include wound healing problems and infection, which have been reported in 11% and 6% of patients postoperatively, respectively.[43,44] Only 1 patient in the present study had wound dehiscence—due to a fall on the knee. There were no issues with routine wound healing or infection in our patients. This is most likely because of our cohort being restricted to patients with hypermobility EDS, as wound healing and skin breakdown are issues commonly seen with other subtypes of EDS.[20] Patients with EDS should be counseled preoperatively about potential complications and surgical risks. From a physician standpoint, it is recommended that multimodal pain management and a multidisciplinary team (genetics, pain management, behavioral therapist/psychiatrist, physical therapist, primary physician, and/or rheumatologist) are involved in management of these patients before planning surgical stabilization procedures.[7]

Articular cartilage injury affecting the patellofemoral joint can be present in up to 95% of patients after patellar dislocation, ranging from minor fissures or cracks to full-thickness chondral defects.[22] Eighteen patients in our cohort required chondroplasty at the time of MPFLR. At revision surgery, 5 patients required patellar chondroplasty, and 2 required subsequent patellar chondroplasty. In patients with EDS, the pathologic collagen can affect all tissues, including cartilage. This can accentuate chondral lesions and can lead to cartilage delamination and progressive wear. There is also an increased association between EDS and rheumatic conditions such as rheumatoid arthritis and ankylosing spondylitis.[26] Joint laxity itself can contribute to premature arthritis.[34] Thus, patients and families need to be counseled about potential knee arthritis and the need for subsequent treatment. Rose et al[29] reported the need for total knee arthroplasty in young patients with EDS, with a mean age of 43.3 years.

Howells and Eldridge[12] reported improved functional outcome scores in patients with hypermobility but to a lesser extent than in controls. Compared with PROs following MPFLR in non-EDS patients, our patients reported inferior outcomes across all PROs (Appendix Table A2).[5,19,21,30] Within our cohort, there was a wide range of PROs, with some patients having significant disability while others were very functional. This reflects the differences in clinical severity in patients with EDS. Such differences are seen even between family members. There were no differences in postoperative PROs when the cohort was divided based on age, laterality (unilateral vs bilateral), graft type, or Beighton score, which is likely because of the smaller sample size in each subgroup (Appendix Table A1). Despite inferior PROs, most patients reported satisfaction at the final follow-up. This may be due to significant improvements compared with preoperative symptoms, the placebo effect of surgery, less overall expectations, better coping mechanisms over years, or other overwhelming conditions that can overshadow their knee complaints.

Limitations

There are several limitations in this study. The retrospective nature of this study has inherent limitations, including recall bias and lack of preoperative PROs. A control group of patients without EDS would have allowed a comparison of the results of the present study, but outcomes of isolated MPFLR in the normal population have been widely published—including a systematic review and meta-analysis.[31] If the subgroup analysis did not show statistically significant differences, it could be due to a smaller sample size and a lack of power in the study to detect such differences. Since the series spanned a decade, some surgical modifications have been made to the MPFLR technique, including a single patellar tunnel and routine use of fluoroscopy to identify the MPFL femoral attachment. Postoperative radiographs were not available for all patients to determine femoral tunnel positioning. Similarly, information related to postoperative physical examination findings—including J-sign, apprehension, or patellar translation—were not available in a consistent manner. Despite these limitations, the present study is the first to report the midterm clinical outcomes of MPFLR in the EDS population. This information should help the physician in patient counseling and surgical planning.

CONCLUSION

Isolated MPFLR restored patellar stability in patients with EDS at the midterm follow-up, although 19.1% had recurrent instability and 38.3% required subsequent surgeries. Failure was more likely in younger patients and those who could touch the palm to the floor with their knees extended. Postoperative PROs were inferior, but patient satisfaction was high.

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APPENDIX

TABLE A1
Comparison of PROs Between Groups Based on Age, Laterality, Beighton Score, and Graft Type[a]

	Pedi IKDC	Kujala	HSS Pedi-FABS	BPII 2.0	KOOS-Pain	KOOS-Symp	KOOS-ADL	KOOS-Sport	KOOS-QOL
Age, y									
≤15	67.7	72.6	5.5	55.1	73.5	69.8	78.9	61.3	55
>15	70.7	77.3	6.3	62.3	78.7	72.6	85.6	68	57.9
P	.68	.53	.76	.45	.54	.69	.34	.54	.76
Laterality									
Unilateral	72.2	80.8	6.3	64.3	81.9	76.2	88.1	71.7	61.5
Bilateral	67.2	71.1	5.6	55	72.2	67.9	78.3	60	53.1
P	.51	.20	.77	.33	.25	.26	.22	.30	.38
Beighton score									
≤7	64.8	70.2	3.9	51.5	71.5	66	78	58.4	50.3
>7	74.9	81.7	8	68.4	82.8	78.2	88.5	73	63.8
P	.19	.14	.10	.08	.20	.10	.21	.21	.16
Graft type									
Allograft	69.1	76.2	5.9	60.5	77.1	72.3	84.7	65.4	57.2
Autograft	69.3	74.1	5.9	57.4	75.3	70.4	80.4	64.1	55.9
P	.97	.78	.99	.75	.83	.80	.58	.91	.89
Autograft type									
Gracilis	67.7	77.7	3	57.2	78.2	68.9	85.3	65.7	56.3
ST	70.4	71.5	8.1	57.6	73.3	71.4	76.9	63	55.6
P	.80	.60	.15	.98	.71	.82	.51	.87	.96

[a]ADL, activities of daily living; BPII, Banff Patellofemoral Instability Instrument; HSS Pedi-FABS, Hospital for Special Surgery Pediatric Functional Activity Brief Scale; KOOS, Knee injury and Osteoarthritis Outcome Score; Pedi-IKDC, pediatric version of the International Knee Documentation Committee; QOL, quality of life; ST, semitendinosus; Symp, symptoms.

TABLE A2
Patient-Reported Outcome Scores[a]

	Pedi IKDC	Kujala	HSS Pedi-FABS	BPII 2.0	KOOS-Pain	KOOS-Symp	KOOS-ADL	KOOS-Sport	KOOS-QOL
Mean	69.2	75	5.9	58.8	76.1	71.2	82.3	64.7	56.5
SD	19.8	20.1	6.7	25.1	22.5	19.4	21	29.4	25
GJH	NR	64.3[12]	NR	66.2[11]	NR	NR	NR	NR	NR
Historical	73.1[21]	85.8[30]	NR	77.8[5]	87.3[19]	88[19]	93.6[19]	81.8[19]	76.8[19]

[a]The best possible score for the HSS Pedi FABS is 30. The best possible score for all other PROs is 100. ADL, activities of daily living; BPII, Banff Patellofemoral Instability Instrument; GJH, generalized joint hyperlaxity; HSS Pedi-FABS, Hospital for Special Surgery Pediatric Functional Activity Brief Scale; KOOS, Knee injury and Osteoarthritis Outcome Score; NR, not reported; Pedi-IKDC, pediatric version of the International Knee Documentation Committee; QOL, quality of life; Symp, symptoms.