Abstract

Background: Patellar instability is a common problem in Down syndrome patients since their childhood. Several treatments have been proposed, but relapses are frequent and not all surgeries are suitable for growing patients. The aim of the present study is to evaluate the clinical and radiographic outcomes of a modified Roux-Goldthwait technique, for the management of patellar instability in children with Down syndrome at minimum 5-year follow-up.

Materials and Methods: 19 patients (23 knees) affected by Down syndrome surgically treated for patellar dislocation, between 2000 and 2012 were included in this study. The mean age of patients was 9.5 years (range 3.7 – 15 years) and had a Dugdale Grade III, IV, and V patellar dislocation. Trochlear groove dysplasia was present in 15 patients. Each patient was clinically evaluated considering relapse rate, pre- and postoperative range of motion (ROM), Kujala score, and modified Lysholm score. Radiographic examination was performed on standard X-ray considering patellar height, trochlear angle, and patellofemoral congruence angle. Results: The mean followup was 134 months (range 62–206 months). No case of relapse of dislocation was registered with an improved ROM (significant for knee extension, \( P < 0.05 \)). The Kujala score showed significant improvement from a mean preoperative value of 39 ± 6.3 to a mean postoperative value of 92.7 ± 3.4 (\( P < 0.05 \)) at final followup such as the modified Lysholm score (from mean preoperative 55.6 ± 6.3 to mean postoperative of 94.2 ± 2.6). Radiographs performed at latest followup showed a tendency to normalization of all the parameters considered, with a restored patellofemoral congruence and trochlear groove shape and without signs of osteoarthritis.

Conclusion: The present study showed that the Roux-Goldthwait procedure is a valid surgical option for the treatment of patellar dislocation in children with Down syndrome.

Keywords: Down syndrome, pediatric knee, patellar dislocation, patellar instability, trisomy 21

Introduction

Chronic patellar instability in skeletally immature population has been widely investigated in the past decade, although its management remains challenging.\(^1\) Different surgical procedures have been described to treat patellar instability with variable outcomes.\(^2\) Patellar instability could be associated with other diseases such as genetic syndromes: The most common is Down syndrome.\(^3\) Due to the joint instability and ligamentous laxity, patellar dislocation is frequent in Down syndrome patients that suffer from limping, pain, and impaired knee range of movement.\(^4\) Patellar dislocation can also be associated with trochlear groove dysplasia, leading if untreated, to patellofemoral arthritis.\(^3\)

Surgical management is necessary, even though there is no consensus on the best surgical technique to use. Considering the skeletal immaturity, the surgical procedure should regard only soft tissues, avoiding damages on the growth plate.\(^5\)

This study reports the results obtained at minimum 5-year followup on a series of patients younger than 16 years, affected by patellar instability and Down syndrome, surgically treated with a modified Roux-Goldthwait procedure.

Materials and Methods

19 patients (23 knees) affected by Down syndrome, treated for patellar dislocation with a modified Roux-Goldthwait surgical procedure, between 2000 and 2012 were included in this retrospective study. There were 11 females and 8 males, the mean age at surgery was 9.5 years.
(range 3.7–15 years); all patients had Dugdale Grade III, IV, and V patellar dislocation [Figure 1]. No patient had undergone prior surgery to the affected knee [Table 1].

**Operative procedure**

All surgeries were performed by a single senior surgeon (P.F.C.). Under general anesthesia, with the patient supine on the operating table and a tourniquet applied to the thigh, a 10 cm medial parapatellar incision was performed from the midpoint of the patella inferiorly to the tibial tuberosity. The lateral and medial retinacula and the patellar tendon were exposed. An extensive soft tissues’ lateral release was performed, taking care to section the lateral retinaculum proximal to the superior pole of the patella. The patellar tendon was divided longitudinally, and the lateral half was detached, as distally as possible, from its insertion on the tibial tubercle, leaving it attached proximally to the patella. With the knee flexed at 30°, the tendon was transferred medially under its intact medial half and sutured with continuous no. 1 absorbable transosseous sutures on the medial side of the tibia, after the incision of the periosteum in that area. The medial retinaculum was finally tensioned. At the end of the procedure, the stability of the patella was evaluated by passively flexing and extending the knee to confirm that it was located satisfactorily in the trochlear groove and by evaluating its lateral displacement at 30° of flexion. The incisions were closed in layers over a drain and routine dressings and bandages, and a cast with 15° knee flexion was applied.

The postoperative protocol consisted of knee fixed casting for 3 weeks; after removal of the cast, a knee splint was applied for further 3 weeks, with 0–30° of motion allowed and then removed for active movement exercises [Figure 2]. After 3 weeks, patients were allowed to progress from partial to full weight bearing. Once a full range of movement was regained, a progressive return to normal activities was allowed with the protection of a patellar brace for other 4 weeks.

All clinical and radiographic evaluations were performed before surgery and were repeated postoperatively at 1 year after surgery and at the final followup. Final followup took place at a mean of 134 months (range 62–206 months) from the index procedure.

![Figure 1: Preoperative X-ray (orthogonal views + merchant view) showing preoperative patellar dislocation in a 10-year-old patient affected by Down syndrome](image)

**Table 1: Patients demographics and preoperative data**

| Patient Number | Sex | Age at operation | Dugdale classification Right knee | Dugdale classification Left knee | Preoperative trochlear groove dysplasia | Preoperative high patella |
|----------------|-----|------------------|-----------------------------------|---------------------------------|----------------------------------------|--------------------------|
| 1              | Female | 12.3             | 3                                 | \                              | +                                      | +                        |
| 2              | Male   | 10.2             | \                                | 3                              | \                                      | \                        |
| 3              | Female | 7.8              | 4                                | \                              | +                                      | +                        |
| 4              | Female | 6.2              | 4                                | 5                              | +                                      | +                        |
| 5              | Female | 9.5              | \                                | \                              | +                                      | +                        |
| 6              | Female | 3.7              | \                                | 4                              | \                                      | \                        |
| 7              | Male   | 11.4             | 3                                | \                              | +                                      | +                        |
| 8              | Male   | 15               | 3                                | \                              | +                                      | \                        |
| 9              | Male   | 9.9              | \                                | 3                              | +                                      | \                        |
| 10             | Female | 12.6             | \                                | 4                              | +                                      | \                        |
| 11             | Male   | 8.1              | 5                                | 5                              | +                                      | +                        |
| 12             | Male   | 9.6              | \                                | 3                              | +                                      | +                        |
| 13             | Female | 6.4              | \                                | 4                              | +                                      | +                        |
| 14             | Female | 8.7              | 3                                | 3                              | \                                      | +                        |
| 15             | Female | 5.4              | \                                | \                              | \                                      | +                        |
| 16             | Female | 6.8              | 3                                | \                              | \                                      | +                        |
| 17             | Male   | 14.1             | \                                | 5                              | +                                      | +                        |
| 18             | Female | 10               | \                                | 3                              | \                                      | \                        |
| 19             | Male   | 13.3             | 3                                | \                              | +                                      | \                        |

+=Present, positive founding, \=Absent, not involved
Each patient was clinically evaluated considering dislocation relapse, pre- and postoperative range of motion (ROM), Kujala score, and modified Lysholm score. All patients were radiographically evaluated in accordance with the radiological instability criteria. Frontal axial and lateral radiographic views in the Merchant position, flexed at 45°, were performed pre- and postoperatively, calculating the height of the patella (in accordance with Caton and Deschamps), the trochlear groove angle, and the patellofemoral congruence angle (angle formed by bisecting the sulcus angle and central patellar ridge, the angle is given a negative value; when lateral, a positive value is assigned).

Patella was considered high when the ratio between the distance from the anterior border of the tibia to the lower center of the joint face of the patella and the measurement of the joint surface was >1.2. The trochlea was considered flat when the trochlear groove angle (sulcus angle) was >150°. A patellofemoral congruence angle >11° was considered positive for lateral subluxation.

All patients had a preoperative patellofemoral congruence angle >11°. Fifteen patients (17 knees) had trochlear groove dysplasia. Seventeen patients (21 knees) had high patella.

The work was conducted in accordance with the Helsinki Declaration of 1975 as revised in 2000 and after the approval from the local Ethical Committee.

The data result from a minimum of three replicated measurements and expressed as a mean ± standard deviation. Comparisons between pre- and postoperative measurements at the last followup were analyzed using paired Student’s t-test; P < 0.05 was considered statistically significant.

Results

All 19 patients (23 knees) included in this study were evaluated at final followup at a mean 134 months after surgery (range 62–206 months). No patient was lost at followup.

We observed an improvement in the postoperative ROM. It results significantly about active knee extension that went from 13.9° ± 4.7° to 4.91° ± 3.8° (P < 0.05).

The Kujala score showed significant and stable improvement from a mean preoperative value of 39.1 ± 4.7 to a mean postoperative value at 1 year of 93.3 ± 4.2 (P < 0.05) and of 92.7 ± 3.4 at final followup.

The modified Lysholm score showed significant improvement, from a mean preoperative value of 55.6 ± 6.3 to a mean postoperative value at 1 year of 94.7 ± 3.4 (P < 0.05). This result was substantially unvaried at last followup (94.2 ± 2.6) [Table 2].

All patients but three were able to perform recreational activities without pain or limping at last followup. Three patients experienced occasionally limping during daily life activities.

Radiographic examination showed a statistically significant improvement (P < 0.05) about postoperative femoral-patellar congruence and trochlear groove dysplasia. A normal patellofemoral congruence angle was obtained in all patients except 2 (3 knees), as well as trochlear groove dysplasia was normalized in all knees except two. High patella correction was not significantly improved [Table 2].

X-rays performed at latest followup showed in all cases no signs of osteoarthritis (OA) even at longest followup (206 months) [Figure 3].

There was no evidence of patellar redislocation episodes referred or clinically and radiographically assessed at any time after surgery.

One superficial wound infection at 1 month after surgery was managed by oral antibiotic therapy. No further
| Follow up (months) | Postoperative episodes of dislocation | Postoperative improvement in knee extension | Mean Improvement in knee flexion | Kujala Preoperative Improvement | Kujala 1 year follow up | Kujala Final follow up | Lysholm Preoperative | Lysholm 1 year follow up | Lysholm Final follow up | Postoperative trochlear groove dysplasia | Postoperative high patella | Postoperative patellofemoral congruence |
|------------------|-------------------------------------|--------------------------------------------|-------------------------------|-----------------------------|--------------------------|------------------------|---------------------|-----------------------|------------------------|--------------------------------|----------------------|---------------------------------|
| 1                | 76                                  | 0                                         | 7                             | 10                          | 39                       | 90                     | 90                  | 47                    | 98                     | 98                              | \                     | \                               |
| 2                | 71                                  | 0                                         | 0                             | 5                           | 41                       | 95                     | 94                  | 52                    | 95                     | 95                              | \                     | \                               |
| 3                | 62                                  | 0                                         | 14                            | 0                           | 35                       | 88                     | 90                  | 55                    | 90                     | 94                              | \                     | +                               |
| 4                | 91                                  | 0                                         | 10                            | 5                           | 34                       | 85                     | 86                  | 45                    | 98                     | 96                              | \                     | +                               |
| 5                | 96                                  | 0                                         | 12.5                          | 2.5                         | 32                       | 94                     | 93                  | 49                    | 89                     | 89                              | \                     | +                               |
| 6                | 67                                  | 0                                         | 12                            | 0                           | 39                       | 90                     | 90                  | 56                    | 95                     | 94                              | \                     | \                               |
| 7                | 195                                 | 0                                         | 10                            | 10                          | 42                       | 98                     | 94                  | 57                    | 96                     | 92                              | \                     | +                               |
| 8                | 199                                 | 0                                         | 5                             | 5                           | 40                       | 98                     | 96                  | 64                    | 94                     | 94                              | +                    | \                               |
| 9                | 185                                 | 0                                         | 8                             | 5                           | 43                       | 92                     | 90                  | 66                    | 95                     | 92                              | \                     | +                               |
| 10               | 206                                 | 0                                         | 10                            | 0                           | 32                       | 94                     | 92                  | 54                    | 92                     | 90                              | \                     | \                               |
| 11               | 182                                 | 0                                         | 15                            | 0                           | 33                       | 98                     | 96                  | 47                    | 100                   | 98                              | \                     | + Abnormal                   |
| 12               | 164                                 | 0                                         | 5                             | 5                           | 37                       | 96                     | 96                  | 58                    | 90                     | 94                              | \                     | +                               |
| 13               | 173                                 | 0                                         | 5                             | 5                           | 45                       | 94                     | 94                  | 61                    | 95                     | 95                              | \                     | \                               |
| 14               | 149                                 | 0                                         | 7.5                           | 5                           | 37                       | 88                     | 91                  | 52                    | 96                     | 96                              | \                     | +                               |
| 15               | 143                                 | 0                                         | 5                             | 0                           | 45                       | 91                     | 93                  | 64                    | 96                     | 95                              | \                     | \                               |
| 16               | 157                                 | 0                                         | 8                             | 0                           | 42                       | 98                     | 98                  | 55                    | 100                   | 98                              | +                    | \                               |
| 17               | 125                                 | 0                                         | 19                            | 5                           | 35                       | 89                     | 87                  | 55                    | 92                     | 92                              | +                    | Abnormal                      |
| 18               | 107                                 | 0                                         | 10                            | 0                           | 43                       | 95                     | 94                  | 54                    | 90                     | 92                              | \                     | \                               |
| 19               | 98                                  | 0                                         | 5                             | 0                           | 48                       | 100                    | 98                  | 65                    | 98                     | 96                              | \                     | \                               |

+=Present, positive founding, -=Absent, not involved
complications were registered and no further surgeries were performed on the operated knee.

Discussion

Management of patellar instability in skeletal immature patients is challenging for the orthopedic surgeon. The treatment of this disease must correct the instability, avoiding damages to the open tibial physis. Tibial tubercle transfer in children could damage the upper tibial physis causing genu recurvatum due to iatrogenic epiphysiodesis.1

Many studies are focused on the management of patellar instability in children and it leads to the development of different surgical techniques involving only soft tissues such as distal realignment procedures (Roux-Goldthwait procedure,12 Galeazzi procedure,13 and patellar tendon transfer,14), proximal realignment procedures15 (VMO advancement), and medial patellar femoral ligament (MPFL) reconstruction.16

Surgical management of such disease is even more challenging in patients affected by ligamentous laxity and joint instability such as in genetic syndromes.

Down syndrome is one of the most common chromosomal abnormalities, consisting of a trisomy of chromosome 21; it is associated with many orthopedic manifestations such as atlanto-occipital and cervical instability, scoliosis, hip instability, slipped capital femoral epiphysis, patellar instability, and foot deformities.13,17

Patellar instability in children affected by Down syndrome leads to pain, limping, and frequent falls that can represent a severe impairment in daily life.12 Surgical intervention should be performed to improve clinical condition and to avoid trochlear dysplasia and early development of patellofemoral OA.18

Treatment options are variable in such patients, and there is a lack of consensus for the most appropriate management. The chosen surgical technique usually reflects surgeon individual preferences and experience.

In literature, there are few studies focusing on the management of patellar instability in Down syndrome. The Green’s quadriceps plasty17 and a modified Roux-Goldthwait12 were used showing good results. Recently, a modified Stanisavljevic procedure was described in a group of patients with fixed or obligatory dislocation showing interesting results either in the subgroup affected by Down syndrome.19

In the present study, we performed a modified Roux-Goldthwait procedure12 consisting of the transfer of the lateral part of the patellar tendon on to the medial side of the tibial tuberosity, associated with the medial retinaculum plication and lateral release.

Our surgical procedure differs from the one described by Bettuzzi et al.3 as they used the Roux-Goldthwait-Campbell surgical technique that consists in using a strip of medial capsule passed through the quadricipital tendon and then returned medially.

The present study showed that the modified Roux-Goldthwait12 technique is effective in treating patellar instability and dislocation in children affected by Down syndrome. Patients had a good clinical and radiographic outcome, demonstrating a statistically significant improvement in knee extension, modified Lysholm14 and Kujala14 scores from the preoperative period to the latest followup, and demonstrating to maintain an adequate patellofemoral congruence and to allow a remodeling of the trochlear groove preventing the development of patellofemoral osteoarthrosis. These results were stable even at long term followup, maintaining unvaried level of clinical outcomes. No recurrences of patellar dislocation were detected and all patients and parents were satisfied with surgery even at long term followup.

Kocon et al.17 reported that good results of eight children affected by Down syndrome after Green’s quadricepsplasty procedure. The MPFL has recently received increased attention as an important stabilizer of the patellofemoral joint and numerous studies in the literature describe effective anatomic repairs or reconstructions of this ligament.1,20,21 Vavken et al.2 have recently shown with a systematic review that reconstruction of MPFL is the most effective management option in recurrent patellar instability in healthy pediatric and adolescent population as this surgical technique restores knee anatomy. It would be interesting to evaluate the long term results of this surgical technique also in syndromic patients, but to the best of our knowledge, no prospective-comparative studies are available in English literature about trisomy 21 patients treated with MPFL reconstruction.22,23

More studies are needed to evaluate if the management of patellar instability in children affected by ligamentous laxity and joint instability, as in syndromic patients, could be the same of other skeletally immature patients. The management of Down patients’ is often very demanding because postoperative management and compliance to treatment in this group is difficult. Moreover, it should be emphasized that children with Down’s syndrome usually have a greater degree of instability, and in our opinion, they should not be considered and managed as standard cases.17

This study adds important findings in the present literature, as it presents a large group of patients with a long term followup in literature surgically treated for patellar instability in Down syndrome.

Limitations of our study include its retrospective nature and the lack of a control group. Another limitation is the lack of more sophisticated imaging assessment of limb-length alignment and rotational and angular profile of the lower limb and knee. We are fully aware that some of the
outcome measurements used in the present study are not validated for children with minor mental disorders, even though the modified Lysholm score and the Kujala score have been previously used in similar patients in pediatric orthopedics.

Conclusion

The present study shows that modified Roux-Goldthwait technique is efficacy in the management of patellar instability and dislocation in patients with Down syndrome. It is a safe and reliable surgical technique with satisfactory clinical and radiographic results at mid and long term followup. Further prospective, comparative study at long term followup is necessary to analyze if it could be considered the gold standard treatment for the analyzed group of patient.

Declaration of patient consent

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

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Conflicts of interest

There are no conflicts of interest.

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