Correction of Bowleg Deformity in Achondroplasia through Combined Bony Realignment and Lateral Collateral Ligament Tightening

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ABSTRACT

Introduction: Achondroplasia is one of the most common osteochondrodysplasias with an incidence of 1 in 26,000 live births. Bowing of lower limbs can cause significant morbidity in this population. The use of the Ilizarov external fixator to tighten collateral ligaments of the knee in children has not been reported in the literature. We report the technique and early results of lateral collateral ligament (LCL) tightening with correction of genu varum in children with achondroplasia.

Materials and methods: A retrospective review of children with achondroplasia presenting with bowleg deformity who were treated by corrective osteotomy and LCL tightening was conducted. Between 1998 and 2003, 12 patients (24 limb segments) underwent this procedure and were included in the study. All patients had grade III LCL laxity preoperatively. Pre- and postoperative anteroposterior standing mechanical axis radiographs were evaluated. The final outcome was graded using the grading system of Paley et al.

Results: All patients had bilateral corrections. The Ilizarov external fixator was used in 10 patients and the Orthofix limb reconstruction system in 2 patients. The bony realignment was achieved through monofocal or bifocal tibial osteotomies. The LCL was tightened in all limb segments using the Paley's type II strategy. The final result was graded as excellent in 20 limb segments and good in 4 limb segments. One patient developed transient common peroneal nerve palsy, four developed grade II pin site infections, and there was premature consolidation of the tibial regenerate in one patient.

Keywords: Achondroplasia, Bowlegs, Genu varum, Ilizarov, Lateral collateral ligament.

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INTRODUCTION

Achondroplasia is one of the most common osteochondrodysplasias with an incidence of 1 in 26,000 live births.^{1,2} It is a form of disproportionate short-limbed rhizomelic dwarfism. Characteristic features include relative macrocephaly, frontal bossing, depressed nasal bridge, bowing of lower limbs, flexion deformity of the elbow, dislocation of the radial head, trident hands, and hyperlordosis. It is caused by mutations in the gene that codes for the fibroblast growth factor receptor 3 (FGFR-3) and is transmitted as a fully penetrant autosomal dominant trait.^{2–5} Most cases are sporadic (85%) and are due to new mutations. At skeletal maturity, the mean adult height is 125–130 cm. There is considerable morbidity and occasional mortality with this disorder such that these patients must be followed closely from birth. Possible problems include stenosis of the foramen magnum and the cervical spinal canal causing cervicomedullary compression with risk of death, central apnea, and neurological dysfunction including paraparesis, respiratory difficulty (from central apnea, a small rib cage and upper airway obstruction), gross motor delay associated with macrocephaly, hydrocephalus, and later-onset neurological problems from lumbar spinal stenosis.^{2,5,6} Hypotonia, genu varum, and joint laxity are common.¹ In addition, there is frequent otitis media, which may be accompanied by hearing loss and delay of speech.³ The effects of short stature and the impact on body image, functional disability, and psychological development are well documented (Figs 1 to 7).

Bowing of the lower limbs can cause significant morbidity in this population. Problems with body image, functional disability, knee pain, and the potential for osteoarthritis in adult life are well

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documented. The incidence of bowlegs in this population is about 50–60%.^{3,7} Surgical management for bowlegs may be necessary in 20–40% of children with achondroplasia. The treatment of bowleg deformity associated with achondroplasia is controversial.^{4,8,9} The effects of brace treatment and fibular epiphysiodesis have not been proven. Surgical management of the bowleg deformity with realignment surgery using osteotomies of the tibia and epiphysiodesis of the fibula has been reported.^{4,10,11} Lateral collateral ligament (LCL) tightening as an adjunct to knee realignment osteotomy has been reported previously in a mixed group of patients of varying diagnoses.¹² The use of the Ilizarov external fixator to tighten knee collateral ligaments has been described for adults but not for, specifically, children with achondroplasia.

Figs 1A and B: Case 1: Preoperative clinical picture

Fig. 2: Case 1: Preoperative planning X-ray

Fig. 3: Closeup view of the knee joints

We report the technique and early results of LCL tightening in addition to bony realignment for genu varum in children with achondroplasia.

MATERIALS AND METHODS

Between 1998 and 2003, 12 patients (24 limb segments) with achondroplasia and genu varum underwent realignment (with or without lengthening) of tibial deformities. There were 11 boys and 1 girl. The median age was 12 years 10 months (range 9–15 years 3 months). All patients had complained of joint instability (wobbly knees) and had chronic laxity of the LCL associated with frontal plane angular deformities of the tibia (with or without femoral involvement). Ligament laxity was graded using the American Medical Association grading system based on clinical examination: grade I: 0–5 mm; grade II: 5–10 mm; and grade III: >10 mm of joint

Fig. 4: Case 1: Postoperative X-ray

opening. All patients had grade III LCL laxity preoperatively and displayed an obvious lateral thrust during the stance phase of gait. Anteroposterior standing mechanical axis radiographs were obtained to evaluate frontal plane alignment and joint orientation pre- and postoperatively. Standing lateral views of both lower limbs were also obtained to assess sagittal plane alignment pre- and postoperatively.

Deformity analysis and correction planning were achieved using the malalignment and malorientation tests. The mechanical axis deviation (MAD)—measured as the distance from the limb mechanical axis to the centre of the knee—was documented before and after surgery. Other measurements included the medial proximal tibial angle (MPTA), the lateral distal femoral angle (LDFA), the joint line convergence angle (JLCA), and the distance of the proximal tip of the fibular head to the knee joint line before and

Fig. 5: Case 1: Follow-up X-ray with regenerate

Fig. 6: Case 1: Consolidation of regenerate

Fig. 7: Case 1: Post-lateral collateral ligament lengthening fibula at anatomical position

after surgery (F1 and F2). The difference between F1 and F2 was calculated as the vertical descent of the fibula.

The final outcome was graded based on a combination of subjective clinical and objective radiographic results using the grading system of Paley et al.¹²

Surgical Technique

All patients had bilateral corrections. The Ilizarov external fixator was used in 10 patients and the Orthofix limb reconstruction system (LRS, Orthofix SRL, Verona) in 2 patients. Bony realignment was achieved using monofocal or bifocal tibial osteotomies. Bifocal correction was performed in 13 segments and monofocal correction in 11 segments. The LCL was tightened in all limb segments using the Paley's type II strategy.¹² The tibia was osteotomised initially

without performing an associated fibular osteotomy. The tibia alone was held in the external fixator proximally (without transfixing the proximal fibula to the tibia), whereas, distal to the osteotomy, both the tibia and fibula were captured by the fixator wires or pins. Correction and lengthening of the tibia in the proximal osteotomy produces a distal movement of the entire fibula relative to tibia, without disruption of the ankle mortise, and a simultaneous tightening of the LCL. Once the lateral ligament has been tightened sufficiently to correct the JLCA, the fibula was then captured proximally in the fixator and a distal fibular osteotomy performed to enable further lengthening. Tightening was deemed complete when the fibular head had descended to the level of the tibial physis or when the JLCA had been overcorrected (Figs 8 to 13).

The postoperative management involved pain relief, pin site care, physiotherapy, occupational therapy, and has been previously described.

RESULTS

Clinical

There were 20 limb segments graded as excellent and 4 as good. The patients were pleased with the outcome of the combined tibial realignment procedure with LCL tightening. The preoperative deformity, instability, and lateral thrust during the stance phase of gait had improved in all patients with the LCL laxity eliminated in all. No clinically significant recurrent instability was noted in any of the patients in the excellent group, but some stretching out of the ligament to a minor degree with time was seen. The median follow-up period was 41.5 months (range 15–96 months). The median fixator time was 160 days (range 95–384 days) and median time to fibular osteotomy was 27.5 days (range 20–42 days) (Table 1).

Radiological

The normal position for the mechanical axis is 1–15 mm medial to the centre of the knee. All patients had medial deviation of the mechanical axis well beyond normal before surgery. The preoperative median MAD was 34.5 mm (28–48 mm) and was corrected to a median of 5 mm (range 0–15 mm) postoperatively

Figs 8A and B: Case 1: Final clinical picture

Figs 9A to C: Case 2: Preoperative clinical picture

Fig. 10: Case 2: Postoperative X-ray

(Table 2). In two children (four limb segments), the value of the postoperative medial MAD was >10 mm due to persisting distal femoral varus. The normal range for the MPTA and LDFA is from 85 to 90° . The tibial varus (MPTA) was corrected to within normal range values; the median MPTA correction was 10° (range 5 – 13°). The normal range for the JLCA is from 0 to 3° medial. The median JLCA correction was 6° (5 – 14°). The median vertical fibular descent was 8 mm (range 5 – 13 mm) of the MAD line when combined with bony correction. A median tibial lengthening of 4 cm (range 3 – 10 cm) was achieved in addition to deformity correction.

Complications

One patient developed a common peroneal nerve palsy toward the end correction that was transient with spontaneous recovery after 3 months. A foot drop splint was used during the recovery

period. Four patients developed grade II pin site infections that were treated with oral antibiotics and rigorous pin site care. Premature consolidation of the tibial regenerate necessitated a repeat osteotomy in one patient. No proximal fibular physal separation was observed in this series (Table 3).

DISCUSSION

Frontal plane malalignment can result in excessive and asymmetrically loaded compartments of the knee joint. During single-leg stance, 75% of the knee joint load passes through the medial tibial plateau in the normally aligned knee. This is due to the medial location of the centre of gravity relative to the knee. Significantly, large medial and lateral MAD are thought to be associated with the possibility of late degenerative changes. This may be a function of the amount of MAD and the load applied

Fig. 11: Case 2: Post-correction

Fig. 12: Case 2: Final X-ray—fibula in anatomical position

Figs 13A to C: Case 2: Final clinical picture

to the knee and therefore affected by the patient's weight and activity level.

Mechanical axis deviation is usually attributed to tibial or femoral deformities or both. It is less often recognised as a result of ligamentous laxity between the tibia and femur. The closer a bony deformity is to the knee, the greater is the MAD produced per degree of angulation. Angulation due to collateral ligament laxity therefore produces the greatest amount of MAD per degree of angulation. Even small degrees of ligamentous laxity can produce large amounts of MAD.

The normal horizontal shear forces on the knee are under 10% bodyweight (5% bodyweight medial shear and 7% bodyweight lateral shear). An increase in horizontal shear forces, amplifying joint instability, may increase the risk for degenerative changes. Laxity of the collateral ligaments leads to increase in the horizontal shear forces on the knee joint. This is perhaps the

strongest argument for collateral ligament retensioning. When correcting lower extremity malalignment, the ligamentous component must be considered. Paley et al. have shown that collateral ligament tightening is a safe adjunctive procedure to bony realignment surgery resulting in a greater degree of correction for the same amount of surgery without significantly increasing the risks.¹²

Bowleg deformity has been observed in 50–60% of children with achondroplasia.^{1,3–5,7} The bowleg deformity is caused by fibular overgrowth relative to the tibia both proximally and distally. This causes altered relationships of the tibia and fibula at the knee and ankle. A previous study has revealed that bowleg deformity in children <6 years is associated with overgrowth of the fibula proximally resulting in proximal tibial varus and LCL laxity. Children >8 years also develop progressive overgrowth of the distal fibula resulting in distal tibial varus.^{2,7}

Table 1: Clinical parameters (preoperative /postoperative)

Pt. no	Age	Sex	Bowleg deformity		LCL laxity		Lateral thrust	
			Preoperative	Postoperative	Preoperative	Postoperative	Preoperative	Postoperative
1	10 years 2 months	M	Yes	No	Grade III	Grade I	Yes	No
2	14 years 4 months	M	Yes	No	Grade III	Grade I	Yes	No
3	14 years 6 months	M	Yes	No	Grade III	Grade I	Yes	No
4	10 years 5 months	F	Yes	No	Grade III	Grade I	Yes	No
5	14 years 1 month	M	Yes	No	Grade III	Grade I	Yes	No
6	14 years 8 months	M	Yes	No	Grade III	Grade I	Yes	No
7	13 years 7 months	M	Yes	No	Grade III	Grade I	Yes	No
8	9 years 11 months	M	Yes	No	Grade III	Grade I	Yes	No
9	15 years 3 months	M	Yes	No	Grade III	Grade I	Yes	No
10	10 years 10 months	M	Yes	No	Grade III	Grade I	Yes	No
11	9 years 1 month	M	Yes	No	Grade III	Grade I	Yes	No
12	12 years 2 months	M	Yes	No	Grade III	Grade I	Yes	No

Table 2: Radiological parameters (preoperative /postoperative)

Pt. no	MAD (medial in mm)		MPTA (°)		JLCA (°)		LDFA (°)	VFD (in mm)
	Preoperative	Postoperative	Preoperative	Postoperative	Preoperative	Postoperative	Preoperative	8
1 R	32	3	79	88	8	2	88	8
1 L	34	0	78	89	8	2	89	8
2 R	30	5	82	90	8	2	91	6
2 L	34	3	80	92	8	2	92	8
3 R	28	2	81	91	9	2	87	8
3 L	30	0	82	90	7	3	89	5
4 R	48	15	80	91	16	2	94	13
4 L	46	12	82	91	14	2	94	12
5 R	40	4	82	89	8	2	91	9
5 L	36	5	80	90	8	3	91	8
6 R	34	13	81	92	9	2	94	7
6 L	35	12	80	91	9	3	94	5
7 R	36	4	79	88	8	2	88	7
7 L	35	3	76	89	8	2	87	9
8 R	32	3	81	86	8	2	92	5
8 L	34	4	80	87	8	2	91	5
9 R	38	5	82	89	7	2	88	7
9 L	36	6	81	88	9	3	89	7
10 R	34	5	82	87	9	2	92	8
10 L	36	6	80	86	8	2	91	6
11 R	38	5	79	89	8	3	89	5
12 R	34	4	78	88	8	3	88	9
12 L	34	3	80	90	8	2	89	9

Bowleg deformity of the lower limbs may remain asymptomatic or can cause significant morbidity in this population. Symptoms, when present, are pain, knee instability, limitation of joint function, a waddling gait, problems with body image, and, rarely, osteoarthritis. The potential for osteoarthritis of the knee in adult life has been observed occasionally in the limited number of natural history studies undertaken in this population.^{1,5,8,10}

Treatment of bow leg deformity remains controversial. The need for surgical correction of some lower limb deformities in achondroplasia is recognised but the specific criteria for treatment have not been established. Brace treatment has been suggested but data to support this treatment are not available

at present. Surgical intervention may be necessary in 10–40% of patients with genu varum. Fibular epiphyseodesis, fibular head resection, tibial and fibular valgus osteotomies, and bony realignment with LCL tightening have been suggested.^{7,8,10,12} The beneficial effects of fibular head resection and fibular epiphyseodesis are not yet proven. Mid-term results of valgus tibial and fibular osteotomies have been reported to be successful in this population.¹¹ Based on his experience, Kopits has suggested that surgical correction be undertaken in patients with achondroplasia who have (1) significant malalignment at any age and (2) asymptomatic malalignment before completion of skeletal maturity.^{3,4}

Table 3: Treatment and outcome parameters

Pt. no	Strategy	Fixator time (days)	FU period (months)	Deformity correction	Tibial lengthening (cm)	Patient satisfied	Complication	Final outcome
1	Bifocal/Ilizarov	95	42	Achieved	3	Yes	None	Excellent
2	Monofocal/Ilizarov	132	90	Achieved	4	Yes	None	Excellent
3	Bifocal/Ilizarov	138	85	Achieved	4	yes	Pin site infection	Excellent
4	Bifocal/Ilizarov	242	30	Persisting DF varus	8	Yes	Pin site infection	Good
5	Bifocal/Ilizarov	105	24	Achieved	3	Yes	None	Excellent
6	Bifocal/Ilizarov	145	24	Persisting DF varus	4	Yes	None	Good
7	Bifocal/Ilizarov	241	66	Achieved	7	Yes	Transient peroneal N palsy	Excellent
8	Bifocal/Ilizarov	146	30	Achieved	4	Yes	None	Excellent
9	Bifocal/Ilizarov	380	41	Achieved	8	Yes	Pin site infection	Excellent
10	Monofocal/Ilizarov	224	15	Achieved	5	Yes	None	Excellent
11	Monofocal/LRS	384	96	Achieved	9	Yes	Pin site infection	Excellent
12	Monofocal/LRS	376	91	Achieved	8	Yes	Premature consolidation tibial regenerate	Excellent

Combined bony realignment with collateral ligament tightening has been shown to provide a greater correction for the same amount of surgery. This technique addresses both the static and dynamic components of the deformity. Collateral ligament tightening reduces the horizontal shear stresses on the knee during single leg loading and may contribute to knee joint preservation. Good results have been reported by Paley et al. at a mean follow-up of 1 year in a mixed group of patients.¹²

CONCLUSION

This is a retrospective study of children with achondroplasia with bowleg deformity treated by bony realignment and LCL tightening. Surgery with uni- or bifocal tibial realignment and a type II collateral ligament tightening was performed. The tibia was lengthened differentially in relation to the fibula by an amount sufficient to allow realignment and LCL tightening. The lateral ligament laxity that had contributed to more than 40% of the MAD in this series was improved to within normal range. Excellent to good results were achieved in all patients without major complications.

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