A Multicenter Longitudinal Study of Intramedullary Rodding in Moderate-Severe Osteogenesis Imperfecta

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INTRODUCTION: Osteogenesis Imperfecta (**OI**) is the most common skeletal dysplasia which predisposes to recurrent **fractures** and **bone deformity** [1] and presents **with wide clinical variability**. The original clinical classification into types I, II, III and IV [2] was modified based on the clinical severity into mild, moderate, severe, and extremely severe [3].

OI require **Multidisciplinary treatment:** medical management, physical therapy and orthopaedic surgery. Surgery goal is twofold: correct long bone deformity and reduce the incidence of fractures [4]. Bone deformities require straightening by **osteotomy** surgery and **intramedullary rodding**. In the last 15 years new rods and modifications of the techniques have been developed to decrease associated trauma [5].

In relation to mobility, it has been postulated that intramedullary rodding is related to the severity of the disease and provides consequences for the ability to walk [6]. Other studies which compare mobility before and after rodding, reveal benefits in ambulation and mobility outcomes [7,8].

Objective: Describe **patterns of rodding** surgery and the **relationship** with mobility and fractures/year in OI types III and IV.

METHODS: Data was collected by the Linked Clinical Research Centers, a network of five centers with significant experience in treating patients with OI [1]. Data included: rodding of femur, tibia and humerus, type of rod, fractures/year, Gillette Functional Assessment Questionnaire (FAQ), Functional Mobility Scale (FMS) and Brief Assessment of Motor Function (BAMF). Relations with mobility outcomes 2 years of age. Analysis focused on Types III and IV.

RESULTS: 558 subjects: 44.6% male, Median Age: 12.4 yrs (0.0 - 67.2 yrs), Follow up: 3.15 yrs (1-5 yrs). OI types III:110, IV:153.

Mean rodded bones/patient: 2.0 in OI type III and 1.67 in type IV. Table 1: % of rodding in each bone, age of rodding. Table 2: types of rods used.

Sequence of rodding: 1st Femur: 58.9%, 1st Tibia: 34.6%.

Simultaneous rodding: **In the last 15 years** 43.1% vs 31.3%. Fracture analysis in Table 3.

Patients started walking before being rodded: 86.8% of type IV and 51.2% of type III.

FAQ Scores: OI type III rodded group showed better results in ($p= 0.01$). No difference between the type IV groups ($p = 0.45$).

FMS (5,50,500 m): No relation with rodding status in OI type III ($p=0.40$, $p=0.46$, $p=0.23$) with better results in OI type IV non-rodded group ($p=0.03$, $p=0.028$, $p<0.01$).

Gillette FAQ Tasks: OI type III rodded group performed better than non-rodded group ($p<0.01$), OI type IV non-rodded group performed better than rodded group ($p<0.01$).

BAMF: better results in OI type III rodded group ($p<0.01$). No differences between groups in OI type IV ($p=0.29$).

DISCUSSION: The prognosis of a child with OI and its relation to rodding surgery are of clinical interest, particularly **to determine when to rod and setting goals for rehabilitation.** The results of this analysis illustrate rodding patterns in moderate and severe OI: type III patients are rodded more frequently, earlier, and in more bones than type IV. In OI type III the rodded group showed better results in most mobility outcomes and lower fracture rate than in the non-rodded group. These **benefits** of surgery were **not observed in OI type IV**. Although this study helps to understand disease related characteristics, the main limitations of this analysis is that the clinical variability within OI types is wide, and we are not comparing mobility outcomes before and after surgery, where additional benefits have been reported [7,8].

SIGNIFICANCE: Analysis of current practice in 5 specialized orthopedics centers will **increase** the **comprehension** of **benefits** and **limitations** associated with OI surgical rodding.

Table 1: Percentage and Age of Rodding in OI Types III and IV

OI Type	N Rodded (%) / Median Age		
	Femur	Tibia	Humerus
Type III	137 (63.7%) / 3.9	117 (54.2%) / 5.3	38 (17.9%) / 8.4
Type IV	182 (59.5%) / 7.6	114 (37.3%) / 8.7	16 (5.2%) / 6.6

Table 2: Types of Intramedullary Rods used.

Rod Type	Femur	Tibia	Humerus
Expanding Intramedullary Rod	80.7%	47.2%	61.4%
Non-Expanding Intramedullary Rod	19.3%	52.8%	38.6%

Table 3: Fractures per Year for OI Types III/IV. Rodded vs Non-Rodded.

	Fractures/Year - Type III				Fractures/Year - Type IV			
	Femur		Tibia		Femur		Tibia	
	Rodded (36)	Non-Rodded (10)	Rodded (19)	Non-Rodded (8)	Rodded (37)	Non-Rodded (14)	Rodded (25)	Non-Rodded (12)
Mean	0.79*	1.31*	0.57*	0.84*	0.87	0.79	0.93	0.81
Min	0.25	0.33	0.25	0.5	0.25	0.25	0.33	0.25
Max	2	4	1	1	2	1.5	2.5	1.5

*Significant difference (level of significance: 0.05)

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