

## **Limb Stabilization In Osteogenesis Imperfecta Patients; Never-ending Battle**

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Osteogenesis imperfecta(OI) is a heterogeneous group of diseases manifesting as congenital bone fragility that leads to frequent bone fractures and development of deformity in the spine and limbs. More than 15 genes have been identified as pathogenic for OI. They are inherited as AD, AR or even XL pattern. Difference in phenotype according to the genotype is currently under investigation, and some genotypes showed their own specific characteristics.

There have been two important steps in the limb stabilization of OI patients – intramedullary rodding and bisphosphonate medication. Bisphosphonate, an osteoclast inhibitor, was proven efficient in decreasing fracture frequency, improving ambulatory status and general well-being of OI patients. However, it can never normalize the bone quality, and surgical stabilization remains indispensable until now. The concept of multiple osteotomy and internal fixation with an intramedullary rod greatly improved clinical outcome of OI patients. As the surgical stabilization needs to be performed from early childhood, elongating rod system fulfils the requirement of growing children. We have devised an telescopic rod system that does not require distal joint arthrotomy nor damage the distal articular cartilage. Even with recent advancement in the design of telescopic rod, series of complications related to either the disease itself or instruments ensue – including failure to telescope, cutting-through, backing-out, and refracture. Revision surgery can be challenging due to difficulty in removal of preexisting rod, distorted intramedullary bony geometry and relatively narrow bone segment at older age. When selecting and implanting an intramedullary rod, revision should always be taken into consideration. Surgeons should be well prepared not only for the initial rodding but also for management of subsequent problems and revision surgery.