Summary
Osteogenesis imperfecta is a hereditary pathology characterized by the osseous fragility, which causes increasing severe deformities in patients. It affects children and it regresses by puberty. We report a particular observation of 16 year old adolescent boy who presented with tardy form of osteogenesis imperfecta. He was treated by intra-medullary nailing. A review of the literature was done.

Key Words: Osteogenesis Imperfecta, Femur, Nailing

Introduction
The Osteogenesis imperfecta (OI) is a group of hereditary disorders characterized mostly by the abnormality of synthesis of collagen type I. The disease is expressed mostly through osseous fragility and deformities. They often present with multiple fractures in childhood, and it normally regresses by puberty. Intra medullary nailing of long bones remains the method of choice for correcting the deformities and preventing fractures.

Case Report
The patient is a 16 year old boy that is not schooling. He is the last of four siblings, and he was admitted to correct deformities of the left femur. There is no similar case in the family.

The history of the disease started at the age of 12 years when the patient presented with frequent fractures of the left femur (3 times). These were treated with Plaster of Paris cast. It progressed to deformity of the femur. Clinical examination revealed small sized, defective and gray set of teeth.

Plain radiography of the left femur showed mid shaft non union (Fig. 1) with mild osteopaenia. The femur had an antero-lateral bow. Calcium phosphate was within the normal range. The treatment consisted of quadruple osteotomy of the femur and fixation with Gross and Kempf intra medullary nail, (Fig. 2), followed by long term rehabilitation.

Our patient was followed up and reexamined ten months later. An excellent outcome was recorded expressed in terms of normal alignment of the femur and absence of fractures.

Discussion
The osteogenesis imperfecta known also as "LOBSTEIN" disease is a genetic disease with dominant hereditary transmission. It is expressed by a reduction of the production of the collagen type I, or by a production of abnormal collagen. Transmutations were localized on the chromosomes 7 and 17. This disease appears in childhood before puberty. The clinical expressions are very polymorphic, associated to variable degrees with: a generalized osteoporosis, bluish sclera, reduced size, ligament laxity, deafness, gray and breakable teeth and cardiopulmonary disorders. The osteoporosis is responsible for the osseous fragility, which accounts for the frequent fractures even following minor accidents (glass bone disease). The most used classification is the modified Sillence classification. Our patient is type IV. No specific
diagnostic test for OI is available. Therefore, differential diagnosis is important. The medical treatment based on biphosphonate has revolutionized the therapy of OI, and seems to be more efficacious in young patients. Some authors recommend orthosis allowing a protective support.

The preferred treatment of fractures and deformities of long bones is intra medullary nailing; this allows the alignment of the fragments, a decreased number of fractures and a better functional result.

The introduction of telescopic nails that has the advantage of elongation with the growth of the bone has revolutionized the treatment of the OI patients. Indeed, the rate of second surgical intervention varies between 20% and 40% while it is more than 50% for the standard nails. However, some mechanical complications such as migration or twisting of the nail and epiphysiodesis must be avoided by developing the ancillaries.

**Conclusion**

Osteogenesis imperfecta is characterized by osseous fragility. Recently, treatment using biphosphonates has improved prognosis in severe forms. There is functional improvement by intra medullary nailing of the long bones. However, it is hoped that genetic therapy will improve spectacularly and possibly eradicate this pathology.

**References**


Fig 2. Post operative stabilization of the left femur with an intra medullary nail.