بِلَادِ النَّاسِ
OUTCOME OF SHISH KABAB OSTEOTOMIES FIXED WITH SIGN NAIL IN OSTEOGENESIS IMPERFECTA.

OUR EXPERIENCE AT GHURKI TRUST TEACHING HOSPITAL, LAHORE, PAKISTAN.
rare and serious disorder of collagen.
make the bones weak and fragile,
results in deformity
being more likely to fracture
The name osteogenesis imperfecta dates to at least 1895.
condition has been found in an ancient Egyptian mummy from 1000 BC.

The Norse king Iva the Boneless may have had this condition, as well.
The earliest studies of it began in 1788 with the Swede Olof Jakob Ekman.

described the condition in his doctoral thesis and mentioned cases of it going back to 1678
In 1831, Edmund Axmann described it in himself and two brothers. Jean Lobstein dealt with it in adults in 1833.

Willem Vrolik did work on the condition in the 1850s.

The idea that the adult and newborn forms were the same came in 1897 with Martin Benno Schmidt.
Genetics

- 90% have an identifiable genetic mutation
  - COL 1A1 and COL 1A2
    - causes abnormal collagen cross-linking via a glycine substitution in the procollagen molecule
- both autosomal dominant and autosomal recessive forms
- can be severe or mild (tarda form)
- **Silence** originally classified into four types however most likely a continuum of disease
- Additional types have been added
- 90% of patients can be grouped into the Silence **Type I and IV**
Type 1
MILD

- COMMONEST
- Fx AT AGE OF 1-2 YEAR
- HEALING GOOD, LESS DEFORMITIES
- DEEP BLUE SCLERA
- DENTINOGENESIS IMPERFECTA
- QUALITY OF LIFE GOOD
TYPE 2
LETHAL

- 5-10 PERCENT
- INTRAUTERINE DEATH AND NEONATAL Fx
- LARGE SKULLS
- SCLERA GREY
- RIBS Fx AND RESPIRATORY FAILURE
- STILL BIRTHS OR DEATH VERY EARLY
- DUE TO DOMNANT MUTATIONS
CLASSIC BUT NOT MOST COMMON
Fx AT TIME OF BIRTH
LARGE SKULLS
MARKED DEFORMITIES AT AGE OF 6 YEARS
SCLERA GREY BECOMING WHITE
DETINOGENESIS IMPERFECTA
MARKED JOINTS LAXITY
POOR LIFE QUALITY
FEW SURVIVE TO ADULTHOOD

TYPE 3
SEVERE DEFORMING
TYPE 4
MODERATELY SEVERE

- UNCOMMON
- FREQUENT Fx EARLY CHILDHOOD
- DEFORMITIES COMMON
- SCLERA PALE, BLUE OR NORMAL
- DENTINOGENESIS IMPERFECTA
- SURVIVAL GOOD
- Thin cortices
- Generalized osteopenia
- Saber shins
- Skull radiographs reveal wormian bones
CLINICAL FEATURES
bone fragility and fractures
  - fractures heal in normal fashion initially but the bone does not remodel
  - can lead to progressive bowing

ligamentous laxity
short stature
scoliosis
codfish vertebrae (compression fx)
basilar invagination
olecranon apophyseal avulsion fx
NON ORTHOPAEDIC MANIFESTATIONS

- blue sclera
- hearing loss
- brownish opalescent teeth *(dentinogenesis imperfecta)*
- wormian skull bones
- increased risk of malignant hyperthermia
OI occurs in about one per 20,000 live births
suggested by the occurrence of bone fractures with little trauma
and the presence of other clinical features.

A skin biopsy can be performed to determine the structure and quantity of type I collagen.
DNA testing can confirm the diagnosis, however, it cannot exclude it because not all mutations causing OI are known and/or tested for.

OI type II is often diagnosed by ultrasound during pregnancy, where already multiple fractures and other characteristic features may be present.
An important differential diagnosis of OI is child abuse, as both may present with multiple fractures in various stages of healing.

- rickets
- osteomalacia and
- other rare skeletal syndromes.
Management of OI is multidisciplinary.

The standard of care includes

- pain management
- therapy input for muscle strength and range of movement
- aids to daily living and mobility as well as
- Surgical management for correction of deformities.
Metal rods can be surgically inserted in the long bones to improve strength, a procedure developed by Harold A. Sofield, MD, at Shriners Hospitals for Children in Chicago.
In 1959, with Edward A. Miller and Sofield wrote a seminal article describing a solution that seemed radical at the time: the placement of stainless steel rods into the intramedullary canals of the long bones to stabilize and strengthen them.
SHISH KEBAB OSTEOTOMY

- requires intramedullary rodding with multiple osteotomies—in lay terms, a shish kebab operation.

- The curved bone is re-broken in multiple places, a rod is inserted down the middle, and the bones are allowed to heal in a straightened position.
STUDY
OBJECTIVE:

- To evaluate the results of shish kabab osteotomies fixed with SIGN NAIL in long bone deformities/fractures of patients suffering from osteogenesis imperfecta.
SETTING AND DURATION:

- Orthopaedics and Spine Centre, Ghurki Trust Teaching Hospital, Lahore, Pakistan from Jan 2014 to March 2016.
METHODOLOGY:

- This study was carried out on 06 patients who presented in OPD or Emergency Department with deformities of long bones.
- Detailed history, examination and investigations including X-ray AP and Lateral view of involved bone were done.
- All these cases were treated with Shish Kabab Osteotomies and SIGN NAIL under spinal or general anaesthesia.
- These cases were followed up for up to 06 months and results of the nail were observed in terms of union, rehabilitation and complications.
RESULTS:

- It was found that 4 (66.7%) were male and 2 (33.3%) were females with a mean age of 11.83±5.24.

- 2 (33.3%) patients having age ≤ 10 years and 4 (66.6%) were between 11-20 years.
The parents of all patients had a first degree consanguineous marriage.

The mean hospital stay were 4.17±1.75 days.

All osteotomies sites heal and after a mean follow up of 06 months, better bone densities, 1 Having recurrence of deformity and no fracture were found.

Only one patients using walkers while remaining need no support for walking.
Case 1; Shamila, 13/F
Immediate post opp.
After 3 months
After 6 months
CASE 2, AHMAD
k. Nailing done else where
After 6 months
After 18 months
CASE 3, Hassan
after 06 months
After 1 year
ANY QUESTION.....