The Surgical Management of Rickets & Osteogenesis Imperfecta

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Rickets

- Inadequate mineralization of growing bone
- Often hypotonic with delayed motor milestones
- Joint thickening esp wrists & knees
- Short for age
- Genu varum / valgum, coxa vara, protrusio
- Pathological fractures
- X-Rays – Loosers zones dt unmineralised osteoid
Rickets

• Wide variation of deformity
• Progression often not predictable BUT majority of deformities correct with medical management
• Indications for surgery
  – Pain
  – Deformity
  – Gait disturbance & activity limitation
• High recurrence rate if not medically controlled
Rickets Management

• Medical management cornerstone of treatment
• Historically
  – Osteoclasis / osteotomies & casting done
• Bracing used briefly
• Be aware of axial / torsional deformities needing correction
Rickets Management

• Sofield & Miller 1950’s – mainly OI but also Rickets
  – Intramedullary fixation preferable to plating
• Acute vs gradual correction – use of Ilizarov external fixators – Paley & Tetsworth
• Multi-apical deformities
• Low energy tissue respecting osteotomies
• Use of growth modulation methods – 8 plates / staples
Rickets
Case One

- Six year old female
- X-Linked hypo-phosphataemic Rickets
- Genu varum both legs
- ALP 589, dropped from 1000
- Bilateral lateral closing wedge osteotomies femurs
Rickets
Case One
Genu Varum
Case One
Internal Tibial Torsion and increased anteversion
Case One
Femoral Osteotomy
Rickets, Case One
Case One
Rickets
Case Two

- 10 year old boy
- 40 degree bilateral genu valgum
- 8 plates inserted bilateral femurs and tibiae
- Corrected and removed after 4 years
LDFA decreased & MPTA increased

On presentation

Four year follow up
Rickets & SCFE

- Slipped capital femoral epiphysis (SCFE)
- More common with renal osteodystrophy
- Slip through metaphyseal side of physis
- Younger age
- Always perform bilateral in situ pinning dt risk of contralateral slip
- AVN more commonly dt steroids than acute unstable slip
- Consider smooth pins to prevent coxa breva
- Beware hardware cut-out and pin penetration dt soft bone
Renal osteodystrophy in a 12 year old boy. Three year follow up, right SCFE, left AVN

Lovell & Winter. Paediatric Orthopaedics. 7th Ed, 151.
Osteogenesis Imperfecta

- Osteogenesis imperfecta
  - Brittle bone disease
  - Genetic basis – spectrum of severity
    - Variants – Bruck’s syndrome
  - Deficiency or abnormality in collagen genes
  - Deformities secondary to multiple fractures
Osteogenesis Imperfecta

- Looser – congenita vs tarda
- Sillence – 4 types
  - Type 5 & 6 – Gloreaux et al
  - Type 7 – Ward et al
Osteogenesis Imperfecta
Natural history

• Based on type
  – Sillence vs newer categorisation
• Site of fracture
• Number of fractures
• Deformity
Osteogenesis Imperfecta
Common Fracture Sites

• Spine – elongated pedicles, kyphoscoliosis, spondylolysis/listhesis
• Humerus - shaft
• Forearm – Monteggia #'s, olecranon avulsion #'s
• Femur – coxa vara, shaft
• Tibia – tibial tubercle avulsion, shaft
• Relatively uncommon
• Often bilateral
  – Up to 70% (Zionts et al)
• Salter Harris type II
• Tension band wiring
• High re-fracture rate if hardware removed
Radial Head Dislocation and Subluxation in Osteogenesis Imperfecta

By Alice Marcdargent Fassier, MD, Frank Rauch, MD, Mehdi Aarabi, MD, Chantal Janelle, MD, FRCS, and François Fassier, MD, FRCS

- 254 Patients, 489 Upper limbs
- Radial head dislocation 44 limbs
- Radial head subluxation 39 limbs
- Overall 17% RCJ malalignment
- 86% Type V
High Prevalence of Coxa Vara in Patients With Severe Osteogenesis Imperfecta

Mehdi Aarabi, MD, Frank Rauch, MD, Reggie C. Hamdy, MD, and François Fassier, MD, FRCSC

- 283 OI patients
  - 94 Type I
  - 90 Type IV
  - 67 Type III
  - 18 Type V
  - 10 Type VI
  - 4 Type VII

- 10% Coxa vara – 55% Type III
- Mean neck shaft angle 99°
Osteogenesis Imperfecta
Pre-operative assessment

• Cardio-vascular assessment
  – Secondary kypho-scoliosis & restrictive lung disease
  – Consider lung function tests
• Hypermetabolic syndrome - history
• Increased bleeding due to extensive surgical exposure – Blood on standby
• Low risk of wound infection
• Choice of surgical implant NB
Osteogenesis Imperfecta
Surgical indications

• New fractures
• Deformities
• Lower limb surgery more frequent than upper limb surgery
Osteogenesis Imperfecta
Intra-operative considerations

• Positioning

• Choice of implant
  – Intramedullary rodding NB vs plate and screws
    • Stress riser
  – Don’t use large diameter rods – causes stress shielding
  – Williams rods vs growing rods (Fassier Duval)

• Sofield Miller procedure
  – Multiple osteotomies

• Valgus osteotomies neck of femur
Osteogenesis Imperfecta
Operative techniques
Sofield-Millar Procedure Femur
Ten patients, 58 long bones
Used a percutaneous technique
Found that > 3 osteotomies resulted in prolonged time to union and thinning of cortices
Osteogenesis Imperfecta
Post-operative care

• Traction
• Derotation bar for femur fractures
• Bisphosphonate recommencement usually once osteotomies have united
Osteogenesis Imperfecta
Potential complications

• Bleeding
• Infection risk low
• Growth and re-fracture at rod end
  — ? Reduced with growing rods
• Rod fracture
• Proximal rod migration
• Extremely fragile bone
Osteogenesis Imperfecta Complications
• 50 Femora analysed
• 14% proximal migration
  – Associated with eccentric rod position distally in epiphysis
  – Special attention with angular residual deformity & persistent osteotomy gap
Management of OI at CHBAH
Oduah G, Firth GB, Pettifor JM, Thandrayen K.
SAOJ Winter 2017

• 78 Patients
• Mean age at presentation 20 months
• 1:1 Male : female
• 26% +ve family history
• 67% presented with a fracture or deformity
• 34 type III, 22 type IV
  – 52% of type IIIs were walking at follow up
• 88% received bisphosphonates
• 66 long bones (49 patients) received IM rodding
  – 77% had revision surgery
Osteogenesis Imperfecta
Case One

- 10 year old girl
- Osteogenesis Imperfecta type III
- PSH
  - Bilateral femoral osteotomies & William’s rods
- Presents with
  - Painful right femur – Distal 1/3 fracture
- Rod Removed and new one inserted
- 2 Osteotomies
- Femur recanalized
- Rod measured
- 25 cm
Overlap osteotomized to minimize tension
Eight Weeks Post-Op
Case Two

- Miss LQ. 12 Years old
- Bruck’s Syndrome
Case Three

- Miss TM, 2 Years old at presentation
- Healed right femur fracture with coxa vara
Case Three

2010

2012
Summary

• Surgical management of Rickets & OI beneficial
• Optimal medical management essential
• Multiple deformities make surgery complicated
• Intramedullary rodding standard
  – Use of elongating rods inherent advantages
• Beware of high complication rate
Thank You