Surgical Management of Fibrous Dysplasia

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1980’s  Few articles or texts
Little experience with F.D.
Tumor experience
NIH contacts
Grateful mother + Internet!
More patients!
Thirty Years

“Accidental” Expert

What Does NOT work
McCune-Albright Syndrome

- precocious puberty
- Café-au-lait
- P.F.D

5 year old female
P.F.D.

- Extent of disease

- Tc 99 bone scan

- “new” areas as child ages
Orthopedic Literature

- Limited
- Monostotic disease
Monostotic Fibrous Dysplasia

- Solitary bone
- Non-aggressive
- Little deformity
- Conventional techniques
Normal Upper Femur X-Ray
Polyostotic Fibrous Dysplasia

- More aggressive
- More deformity
Polyostotic Fibrous Dysplasia

- Thin, weak bones
- Early deformity
- New techniques
Long List of Procedures That Do NOT Work

- Do NOT use patient’s own bone graft
- Do NOT use cancellous graft
- Do NOT try to remove all of the disease
- Do NOT allow the deformity to get severe
- Do NOT use plates or screws **
- Do NOT expect a single operation to solve the problem
DEFORMITY

*Can NOT be cured

*Can be managed
Indications for Surgery

- Control of weight bearing bone pain
- Reverse deformity *
- Diminish fracture frequency
Surgery for Weight Bearing Bone Pain:

No surgery until maximum medical management:

Ricketts Management

Bisphosphonates
Surgery

In extensive disease surgery may be needed as often as every 18 months !!!
Goals

- Early weight bearing
- Avoid casting and bed rest
What have I learned?

- **Less** bone grafting *
- **More** intramedullary rods
- **Early** surgery for upper femur deformity!
Do Not Do Nothing !!!

Age 3

Age 12
Children’s Bones

- Small
- Open physes
Devices

- Wrong bone
- Different insertion techniques
New Devices!!

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Surgical Technique
Hands and Feet

- Rarely need surgery
- Grafting for repeated fractures
Tibias

- Less deformity
- Fractures
- Pain with weight bearing
- Manage with Intramedullary rods
- Inventive!!!!!
Surgery for the tibia is much more effective than surgery for the femur.
Mature patient = adult device
Normal Upper Femur X-Ray
Femur

- Most frequent involvement
- Earliest deformity
- Most difficult reconstruction
Femoral Neck

- Normal angle 135 degrees
- Fibrous Dysplasia causes angle to decrease
- Reversing deformity is difficult
- Operate early!
Femoral Neck

- Consider surgery when Neck-Shaft Angle is 110 to 120 degrees
- Use intramedullary devices when available
Femoral Neck
Must be “inventive” for small children

Pre-op
Post-op
3 yrs Post-op
Femoral Neck

- **Flexible rods:** No support for the femoral neck!
- **Fassier-Duval rods:**
  no support for the femoral neck!
Femoral Neck

- K-wires penetrate the weak bone of the neck
Femoral Neck

- Post op valgus osteotomy
Femoral Neck

- Post op valgus osteotomy on the right
Femoral Neck

- Most recent follow up
Upper Extremities

- Seldom require surgery
- Casting
- Small intramedullary rods if needed
Upper Extremities
Beware!

- Aneurysmal bone cyst
- Expansile liquid filled cyst in previous Fibrous Dysplasia
- Looks like Fibrous Dysplasia on x-ray
- M.R.I.
Suspect Aneurysmal bone cyst in older child with increased pain
Late Reconstructions

- Never very good
- Avoid by doing early surgery!
- “Salvage” using massive allografts and artificial joints
- Risk of Infection!
Case 1

4 year old female
Case 1

July 2008 (age 8)  January 2011 (age 11)
Case 1

October 2009  (age 9)
Case 1

February 2011

April 2011
Case 1

October 2011

November 2012
Case 1

March 2013
Case 1

March 2013
Case 2

October 2009 (age 12)

December 2009 (age 12)
Case 2

March 2010 (age 13)

January 2011 (age 14)
Case 2

August 2011

September 2011

October 2011
Case 3

- 22 year old female
- Severe pelvic, femoral, and tibial disease
- Mild pain
- Ambulatory with crutch
- No surgery
Case 3

- Crutch = upper extremity weight bearing
- Pain
- IM rods
Case 4
Case 5

Age 5
Final Thoughts

- Surgery will NEVER cure Fibrous Dysplasia
- Genetic/Basic science research is key
- Seldom is surgery recommended too early
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