HEALTHY LIMBS, FEET & SPINE
TOPICS

Spine issues
Dural Ectasia
Hips
I. MAIN HEALTH CONCERNS IN MFS

- Heart
- Sight
- Back
- Fatigue

Rao, Sponseller 2016
MAIN SOURCES OF SKELETAL PAIN
-FURTHER RESEARCH NEEDED ON THESE

Back
Hip
Neck
Knee
Shoulder
Feet

Mean Values for Experienced Pain

6
5
4
3
2
1
II. SPINE ISSUES

Scoliosis
Kyphosis
Slipping (Spondylolisthesis)
Scoliosis in Marfan Syndrome

Scoliosis affects 2/3 of MF
Does not always worsen
10% need brace or surgery
Curves increase faster
Increased back pain

97°
WILL BRACE HELP SCOLIOSIS?

Straighten curve? – No

“Hold” it? – Maybe

Results of bracing:
- 20% successes
- 80% failures

Same principle for all connective tissue disorders!!
BRACE RECOMMENDATIONS

Consider brace at earlier point
- 15-20° curves in young children

Avoid in “low-yield” situations
- Bigger curves, reluctant kids
INFANTILE MARFAN SYNDROME

Requiring surgery before age 9
Fusion arrests trunk growth
Solution: “Growing” rods (internal brace)
10 Marfan children
- 80° curves
- 10 cm length in 5 years
- No infections
MAGNETICALLY CONTROLLED GROWING RODS ("MAGEC")

If No need for MRI
ADOLESCENT SCOLIOSIS SURGERY: MFS VERSUS IDIOPATHIC (TYPICAL)

Multicenter, Case-Control

34 Marfan patients

68 AIS patients

- Matched 1:2 for age, gender and degree of major deformity
RESULTS

Marfan vs. AIS:
- More Rod Complications (3 vs 1, \( p = 0.007 \))
- More Reoperations (8 vs 0, \( p = 0.01 \))
- More Intraoperative CSF Leaks (3 vs 0, \( p = 0.01 \))
- Progression of Unfused Thoracic Curves (3 vs 1, \( p = 0.107 \))
SPONDYLOLISTHESIS

Not more common, but more likely to slip
L4-5, L5-S1
Grades 2-5
Posterior fusion only
BONY CHANGES

Pedicles thin
Lamina thin
Vertebrae scalloped
IIC. DURAL ECTASIA

Present in over 50% of patients
- LDS similar

Visual criteria:
- dural diameter S1 > L4
- scalloping > 4 mm
- anterior meningocele

What is Dura?
- Tough tube protecting spinal cord
- Contains clear fluid - CSF
HOW DOES DURAL ECTASIA DEVELOP?

- Spinal Fluid pulsations
- Dilatation of the weak dura
- Scalloping/erosion of the vertebral body
- Signaling effects?
DOES DURAL ECTASIA CAUSE PAIN?
YES, BUT...

Case-control study 30-50 y/o
- DE greater in pain group
- But not all with DE had pain!

Possible reasons for pain:
- pressure on bone, soft tissues
- folding of dura or nerve roots

Treatment?
SIGNS/SYMPTOMS ASSOCIATED WITH DURAL ECTASIA

Very Low Back pain (91%)

Headaches (77%)

Leg pain (64%)

Pelvic & Perineal pain (30%)
DOES DURAL ECTASIA GET WORSE?

11 yr follow up

12 patients
- repeat MRI/CT/pain score
- age 52y/o (42-60)
60Y/O WOMAN WITH SEVERE DE (ANTERIOR MENINGOCELE)
48Y/O MALE WITH MODERATE DE
RESULTS

No statistically significant difference in:
- dural ectasia size noted after 11 yrs
- Symptoms (pain scores)
IV. HIPS
NEONATAL HIP DISLOCATION IN MARFAN

Incidence about 2 %

Pavlik Harness not successful

Surprisingly good response (3/4) to closed reduction and hip spica cast
HIPS
“PROTRUSIO”

Deep hip socket
- Center-edge angle
- Medial displacement

Bone more plastic under load
- With growth
- Mechanism:
  - Activation of osteoclasts?

Are there other reasons for hip degeneration?
RESULTS- PROTRUSIO STUDY

Pain only present in ~5% of 300 hips
- Role of prophylactic surgery seems limited

Few people needed hip replacement
Hip replacement has good results
UNANSWERED QUESTIONS

Mechanism for osteopenia/osteoporosis
Pathogenesis of dysmorphic changes
Mechanism/treatment of
- Skeletal pain,
- fatigue,
- disability