Speaker Name: Luciano Dias, MD; Vineeta T. Swaroop, MD; Rachel M. Thompson, MD

Disclosure of Relevant Financial Relationships
I have no financial relationships to disclose.

Disclosure of Off-Label and/or Investigative Uses:
I will not discuss off-label use and/or investigational use in my presentation.

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**CONTROVERSIES IN ORTHOPAEDIC MANAGEMENT OF PATIENTS WITH MYELOMENINGOCELE**

**Introduction**
Luciano Dias, MD

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**SURVIVAL RATE**
- 1950-10%
- Presently 75% can expect to reach early adult years
- Highest mortality – first year of life

**INCIDENCE**
- From 1983 to 1990 - 4.6 per 10,000
- Gradual decrease
- 2 main factors
  1. Abortion
  2. Folic Acid

**INCIDENCE**
- After folic acid mandate
  - 1.9 per 10,000 live birth

**FMS**
- Functional Mobility Scale
  - FMS
  - Ability to walk 3 specific distances
  - 5 / 50 / 500 meters
**FUNCTIONAL MOBILITY SCALE (FMS)**


**FUNCTIONAL MOTOR LEVEL**

- High Lumbar/Thoracic
- Low Lumbar
- High sacral
- Low sacral

**ORTHOPEDIC CARE**

**START AT BIRTH**

**PHYSICAL EXAMINATION**

**MANUAL MUSCLE TEST**

**SPINE/PELVIS XRAYS**

**HIGH LEVEL**

- No Quadriceps
- MAY HAVE HIP FLEXORS
- RGO/HKAFO
- PARAPODIUM if seating balance is poor
- FMS 2/2/1 when < age of 10
- Most will stop walking by 11 to 13 years of age
- Obesity is common
- Adult-99% wheelchair mobility (FMS 1/1/1)

**HIGH LEVEL**

- FMS 3/3/1

**FMS 2/2/1**
### MOST COMMON DEFORMITIES

- HIP FLEXION CONTRACTURES
- KNEE FLEXION CONTRACTURES
- EQUINUS

### ADULT PROBLEMS

- LOWER EXTREMITY EDEMA
- PRESSURE SORES ISCHIUM

### LOW LUMBAR LEVEL

- HIP FLEXORS - STRONG
- KNEE EXTENSOR - STRONG
- MEDIAL HAMS - STRONG
- GLUTEUS MEDIUS - 2 OR LESS
- GLUTEUS MAXIMUS - 2 OR LESS
- GASTROSOLEUS - 0

### LOW LUMBAR

- Walk with AFO and Crutches at an average walking velocity 60 % of expected
- YOUNG - FMS 3/3/3
- 80% retain walking ability as adult
- FMS 3/3/1
- WHEN OBSESE
- FMS 3/1/1

### LOW LUMBAR

- FMS 3/3/3
- FMS 3/3/1
LOW LUMBAR

• NO SHUNT
• FMS 5/5/3

UNCOMMON

MOST COMMON PROBLEMS

• KNEE FLEXION CONTRACTURE
• CROUCH GAIT
• EXTERNAL TIBIA TORSION
• VALGUS KNEE STRESS

MOST COMMON PROBLEMS

• ANKLE VALGUS
• HINDFOOT VALGUS
• HIP CONTRACTURES
  • HIP DISLOCATION DOES NOT AFFECT THEIR WALKING ABILITY

ADULT PROBLEMS

• SWELING LOWER EXTREMITY
• PRESSURE SORE ANKLE/FOOT
• OBESITY CAN AFFECT THEIR MOBILITY

HIGH SACRAL

• Weak Gluteus Medius and Maximus (2 or+)
• Gastrosoleus strength < 3
• AFO(solid)
• Gluteus Lurch
• FMS 5/5/5 or 5/5/3 or 6/6/6
• Walking velocity 75%
• 94% retain walking ability as adult

HIGH SACRAL; WITH AFO

FMS 5/5/5
MOST COMMON PROBLEMS

- CROUCH GAIT
- KNEE FLEXION CONTRACTURE
- EXTERNAL TIBIAL TORSION
- ROTATIONAL DEFORMITIES FEMUR
- HIP SUBLUXATION

MOST COMMON PROBLEMS

- HIND FOOT VALGUS
- HINDFOOT VARUS
- CAVOVARUS FOOT

LOW SACRAL LEVEL

LOW SACRAL

Less than 5%
- Strong gluteus medius and maximus >3
- Strong gastrosoleus 3 or >
- Do not need AFO
- May need SMO
- No gluteus lurch
- Normal gait
- FMS 6/6/6

FUNCTIONAL COMPARISON BETWEEN SHUNT AND NO SHUNT PATIENTS WITH MYELOMENINGOCELE

BATTIBUGLI

Gait Analysis Parameters

<table>
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<tr>
<th>Linear Parameters</th>
<th>NO SHUNT</th>
<th>SHUNT</th>
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<tr>
<td>Velocity</td>
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<td>76.4</td>
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<tr>
<td>Cadence</td>
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<td>86</td>
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<tr>
<td>Stride Length</td>
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<td>88.4</td>
</tr>
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</table>

Group 3

P- 0.0009 VELOCITY

P- 0.003 STRIDE LENGTH

FMS

FUNCTIONAL MOBILITY SCALE

- 500

HAVING NO SHUNT WAS SIGNIFICANTLY ASSOCIATED WITH
- HIGHER
- FMS 500
- P- 0.01

FMS 6 6 6

SACRAL LEVEL

- Group 1 - No Shunt 71.4%
- Group 2 - Shunt 54.9%
FUNCTIONAL COMPARISON BETWEEN SHUNT AND NO SHUNT PATIENTS WITH MYELOMENINGOCELE

Conclusion

The functional mobility of no shunt patients, was found to be better in our MM study population in relation of the linear gait parameters and Functional Mobility Scale – FMS

Neurosurgeons should be aware of these differences and develop strict guidelines to when insert a shunt

HYDROCEPHALUS

- 1985
  - A REVIEW OF 200 CHILDREN CLOSED AT CMH
    - 92% SHUNT
    - 2010
    - 65%
  - “THE BEST SHUNT IS NO SHUNT”

CONTROVERSIES IN ORTHOPAEDIC MANAGEMENT OF PATIENTS WITH MYELOMENINGOCELE

Hip subluxation/ dislocation
Vineeta T. Swaroop, MD

HIP INSTABILITY

- Affects up to 50% spina bifida patients
- Common but complicated problem
- Treatment remains controversial

HIP INSTABILITY

- 1960s and 1970s
  - Aggressive treatment approach for ALL patients
  - Transfer of iliopsoas tendon
  - External oblique transfer/ varus osteotomy
  - Goal = anatomic reduction of hip
- “Successful” result based on anatomic/ radiographic results
- Little regard for functional consequences
- Concern: did hip reduction lead to ↓ ROM, complications???
HIP INSTABILITY

- Attempted surgical reduction:
  - Multiple studies → high complication rate
  - Worsened neurological deficit, loss of motion, pathologic fractures

- Sherk DMCN 1991:
  - Compared op to non-op treatment
  - Surgical group: 36% ↓ ambulatory capacity

- Bazih JPO 1981:
  - 74 patients with surgical treatment
  - 45% had redislocation
  - Conclusion: surgery had no benefit for ambulation

MORE POOR OUTCOMES

After attempted surgical reduction:

- Sherk and Ames Clin Orthop 1978
  - 53% redislocation rate
  - 11% worsening neurological deficit
  - Pathologic fractures common

- Feiwell JBJS 1978
  - 29% loss of motion
  - 17% pathologic fractures

HIP DISPLACEMENT

Radical change in goals of treatment:

- Previously, goal = anatomic location of hip joint
  - Iliopsoas transfer, external oblique transfer, VDRO
  - Many studies have shown poor functional outcomes

- Now, goal = maximum function, emphasis on gait
  - ‘A level pelvis and a good range of hip motion are more important for function than reduction of the hips. The goal of treatment should be maximum function, not radiographic reduction of the dislocated hips’ Feiwell 1978

HIP INSTABILITY

- General consensus in literature: Ambulatory ability does not depend on status of the hip but rather
  - most important factor is level of functional involvement
  - Barden JBJS, Sherk CORR, Feiwell JBJS, Sherk DMCN, Feiwell CORR

- Preserving muscle strength of iliopsoas and quadriceps is more relevant than status of hip joint in terms of potential for continued ambulation in adulthood

The Effect of Hip Reduction on Function in Patients with Myelomeningocele

1978: Feiwell

- Compared functional results:
  - 35 – surgical hip reduction
  - 41 – no operative treatment

- Surgical group: 40% redislocation rate
  - No improvement in ROM, ambulation
  - No ↓ pain ↓ or need for bracing

- Gait function depends on level pelvis and adequate ROM rather than anatomic joint reduction!
HIP SUBLUXATION / DISLOCATION: THORACIC AND HIGH-LUMBAR LEVELS

- Stability of hip joint has little clinical effect on function
- Treatment limited to contracture release to allow:
  - Proper sitting posture
  - Perineal care
  - Facilitate use of orthoses for ambulation, if appropriate

→ NO CONVINCING EVIDENCE TO SUPPORT HIP REDUCTION IN THIS GROUP

LOW-LUMBAR LEVEL: UNILATERAL DISLOCATION

- Indications for hip reduction?
- 3D Gait analysis study:
  - Examined influence of unilateral hip dislocation on gait
  - 20 patients (low-lumbar)
  - Community ambulators with solid AFOs + crutches

Gabrieli et al J Pediatr Orthop 2003

GAIT PARAMETERS

GROUP I:
- No contracture or symmetric contracture
- 70% patients – symmetric gait pattern (<10° difference btw unstable hip and contralateral side)
- No difference in stride length, cadence, walking velocity or LLD between groups

GROUP II
- Unilateral contracture
- 20% patients – symmetric gait pattern

LOW-LUMBAR LEVEL: UNILATERAL DISLOCATION

GROUP I:
- 10 patients
- No contracture or symmetric contracture

GROUP II:
- 10 patients
- Unilateral hip flexion and/or adduction contracture

Pelvic and hip kinematics assessed to determine symmetry of motion between involved and non-involved side during ambulation:
- Pelvic obliquity, pelvic tilt, pelvic rotation, hip rotation, hip ab/adduction, hip flexion/extension

GROUP I: CHARACTERISTIC PATIENT

Pelvic Obliquity

Subluxed Side in Red

% Gait Cycle

Degree

0 10 20 30 40 50 60 70 80 90 100
**GROUP I: CHARACTERISTIC PATIENT**

- **Hip Abduction/Adduction**

  - Subluxed Side in Red

**GROUP II: CHARACTERISTIC PATIENT**

- **L adduction contracture**

- **GAIT PARAMETERS - UNILATERAL DISLOCATION**

  - In both groups, walking speed = 60% of normal
  - Corresponds to velocity of low-lumbar patients without hip dislocations
  - **Hence, unilateral hip instability does not influence walking velocity**

**LOW-LUMBAR LEVEL: UNILATERAL DISLOCATION**

- Conclusions:
  - Gait symmetry corresponds to absence of hip contractures or bilateral symmetrical hip contractures
  - NO RELATION to presence of hip dislocation
  - **Reduction of hip is unnecessary!**

**LOW-LUMBAR LEVEL: UNILATERAL DISLOCATION**

- Conclusions:
  - Challenges efficacy of surgery to relocate hip
  - Since hip instability in these patients has minimal effect on gait symmetry
  - If contractures are causing gait asymmetry
  - **Address contractures surgically (Treat the FUNCTIONAL deformity)**
  - Gait symmetry will likely be restored
HIP SUBLUXATION /DISLOCATION: SACRAL LEVEL

- Relatively rare BUT challenging treatment dilemma
- In contrast to low-lumbar patients who walk with crutches,...
- In sacral level patient who walks with no support:
- Hip instability can lead to lever arm dysfunction
  - ↑ Trunk – pelvic lurch due to loss of fulcrum
  - ↑ Pelvic obliquity
  - ↑ Gait asymmetry
- Functional abductor strength can be compromised
- Should sacral level patients be considered for surgical relocation???

HIP SUBLUXATION /DISLOCATION: SACRAL LEVEL

- Factors to consider:
  - Prospect for independent ambulation thru adulthood?
  - Magnitude of asymmetry during gait?
  - Joint integrity?
- Careful consideration for surgical reduction in this group to maintain independent ambulation as adult
- Further studies necessary to assess results of surgical treatment

HIP DISPLACEMENT – SACRAL LEVEL

- Muscle weakness – gluteus
- Tethered cord
- We are finding more and more subluxated hips in these patients
  - 6 patients in past 36 months
  - Looking at series with pre- and post-op GA
- Need for screening AP pelvis in sacral level patients?

SACRAL LEVEL

- Treat all cases???
- One stage hip reconstruction:
  - Adductor myotomy
  - Varus derotational osteotomy of proximal femur
  - Bilateral?
  - Open reduction/ capsulotomy
  - Acetabuloplasty

ONE-STAGE HIP RECONSTRUCTION

- Soft-tissue lengthening
- Varus derotational osteotomy of proximal femur
  - Closing wedge osteotomy
  - Blade plate for fixation
  - Femur shortening if needed
  - Goal – Neck shaft angle 110-120°, anteversion 10-20°
- Capsulotomy
- Acetabuloplasty

CONCLUSION

- Available literature supports LEVEL OF NEURAL DEFICIT as most important predictor of ambulatory ability
- Most authors agree: extensive surgery to reduce hip dislocations is not indicated
- Treatment goals = level pelvis, free motion of the hips
- Recommended surgical treatment = contracture release when necessary
TO REDUCE OR NOT TO REDUCE THE HIP – CURRENT RECOMMENDATIONS

- Thoracic/High-lumbar/ Low-lumbar level
  - If contracture exists, causing asymmetrical gait:
    - Treatment with soft tissue release indicated to improve gait
    - No attempt should be made to reduce hip joint

- Sacral level
  - If dislocation present in a child who walks with no support:
    - Consider possibility of tethered cord
    - Surgical relocation indicated to correct lever arm dysfunction
    - Soft tissue release, open reduction, VDRO, pelvic osteotomy

HIP CONTRACTURES

- Can affect function more than subluxation/dislocation
- Untreated →
  - Pelvic obliquity
  - Compensatory spinal deformity
- Gait analysis:
  - Unilateral hip flexion/adduction contracture →
  - ↑ pelvic obliquity →
  - Asymmetric gait/ compensatory scoliosis

HIP CONTRACTURES: ETIOLOGY

- Muscle imbalance
  - E.g. low-lumbar level: lacks normal strength in gluteals → relatively greater strength in hip flexors/adductors leads to hip deformity
  - Type/severity of contracture depends on degree of muscle imbalance present

- Positioning
  - Especially in high levels of involvement – rely on wheelchair for mobility

- Spasticity
  - Tethered cord syndrome

HIP FLEXION CONTRACTURE

- Most common in low lumbar level
- Rare in sacral level
- ↓ Velocity
- ↑ Upper extremity demand
- ↑ Anterior pelvic tilt

Physical exam:

- Thomas test

- Gait:
  - Increased hip flexion during mid-stance
  - Increased anterior pelvic tilt
    - Forward lean of upper body
    - Increased lumbar lordosis
First 2 years of life: hip flexion deformity tends to decrease
→ Treatment rarely indicated in this group

> 2 years of age:
Specific treatment recommendations based on functional level of involvement

HIP FLEXION CONTRACTURE: TREATMENT

TREATMENT: THORACIC, HIGH-LUMBAR

- Flexion contracture < 30–40°
  - Often tolerated if does not interfere w/ orthotic use/ambulation
- Flexion contracture > 30–40°
  - If patient attempts to walk w/ RGO
    - Very short stride length
    - Increased lumbar lordosis
- Treatment indicated:
  - Facilitate use of orthosis for standing/ walking
  - Provide adequate ROM to sit comfortably in wheelchair, lie supine in bed

TREATMENT: THORACIC, HIGH-LUMBAR

SURGICAL TECHNIQUE

- Illofemoral approach
- Start with:
  - Tensor fascia lata
  - Sartorius
  - Rectus femoris
  - Iliopsoas
- If needed, anterior capsulotomy of hip joint

TREATMENT: THORACIC, HIGH-LUMBAR

Post-operative management:
- Short period of full-time use of total body splint (~2 weeks)
- Followed by night-time bracing + aggressive physical therapy

TREATMENT: THORACIC, HIGH-LUMBAR

- Very severe cases: contracture > 60°
- If pressure sores are a problem and soft tissue release alone is not enough:
  - Hip flexor lengthening +
  - Proximal femur extension osteotomy
- AO blade plate for fixation

TREATMENT: LOW-LUMBAR, SACRAL

- Hip flexion contractures can cause major functional impairment
- In patient who walks with AFOs + crutches
  - HFC > 20°:
    - can cause significant anterior pelvic tilt
    - decreased walking velocity
    - increased demand on upper extremities
    - greater energy cost
When surgery is indicated → must take care to preserve hip flexor power

- Release rectus femoris and TFL
- Detach sartorius from ASIS, reattach to AIIS
- IF iliopsoas lengthening is necessary,
  - Do intramuscular lengthening above the pelvic brim
  - NEVER release distally in ambulatory patients!
  - Loss of hip power, inability to forcibly flex hip vs. gravity

Surgical treatment indicated:

- When contracture interferes with function

Adductor longus and gracilis
- Myotomy

Part of adductor brevis if needed
- Protect obturator nerve

Goal = at least 60° abduction

Post-operative management:

- Abduction wedge full-time for short period (~12 days)
- Followed by night-time wedge + aggressive physical therapy

Severe recalcitrant cases:

- Subtrochanteric valgus osteotomy of the proximal femur
  - May be necessary to achieve sufficient abduction to improve pelvic obliquity

Severe stiffness of hip joint

- Patients who have had attempted surgical treatment
  - Major problem
HIP STIFFNESS

- One option: Castle procedure
  - Resection of proximal femur
  - Capsular flap closed over acetabulum

- Goal = improved ROM

- Disadvantages:
  - Need for post-op traction
  - High risk of heterotopic ossification

REFERENCES


OTHER OPTION?

- McHale osteotomy =
  - Femoral head resection + proximal femur valgus osteotomy

- In patients with CP:
  - Allows good ROM
  - Improves sitting ability
  - Eases perineal care

REFERENCES


CONTROVERSIES IN ORTHOPAEDIC MANAGEMENT OF PATIENTS WITH MYELOMENINGOCELE

Knee- crouch gait
Luciano Dias, MD

INTRODUCTION

- KNEE FLEXION CONTRACTURE
- COMMON IN SPINA BIFIDA
- LOW LUMBAR/SACRAL LEVEL
- INCREASE OXYGEN COST
- SURGICAL TREATMENT
- FLEXION CONTRACTURE > 15 TO 20 DEGREES
CROUCH GAIT

- NO GASTROSOLEUS
- WEAK GLUTEUS
- NORMAL QUADRICEPS

LOW LUMBAR LEVEL

- NO GASROSOLEUS
- NO GLUTEUS
- NL QUADRICEPS
- NL HAMSTRINGS

SACRAL LEVEL

- NO GASTROSOLEUS
- WEAK GLUTEUS
- NORMAL QUADRICEPS

CROUCH GAIT

- These patterns of muscle weakness lead to a gradual increase of knee flexion during the stance phase of gait which, in turn, can lead to the development of a gradual contracture of the knee capsule and hamstrings, seen usually around the age of 10

CROUCH GAIT

- INCREASE OXIGEN COST
- INCREASE OXIGEN CONSUMPTION
- DIRECT RELATION WITH THE AMOUNT OF KNEE FLEXION DEFORMITY
---

**SURGICAL PROCEDURE**
- **PATIENT IN PRONE**
- **TRANSVERSE INCISION 1 CM ABOVE THE CREASE**
- **SELECTIVE LENGTHENING MEDIAL HAMS AND BICEPS**
- **THE MEDIAL AND LATERAL ORIGINS OF THE GASTROCNEMIUS ARE FREED FROM THE RESPECTIVE FEMORAL CONDYLES**

---

**INCISION**

---

**INCISION**

---

**SURGICAL PROCEDURE**
- **THE POSTERIOR KNEE CAPSULE IS EXPOSED**
- **POSTERIOR CAPSULECTOMY**
- **POSTERIOR CRUCIATE LIGAMENT IS LEFT INTACT**
  - **SKIN CLOSURE**: INTERRUPTED SUTURE
  - **LONG LEG CAST**
  - **KNEE IN FULL EXTENSION**

---

**POST OP**
- **IMMOBILIZATION**: 3 WEEKS
- **IF FULL EXTENSION NOT ACHIEVED AT THE TIME OF SURGERY**
  - **CAST CHANGE IN ONE WEEK**
  - **AFTER CAST REMOVAL**
    - **AFO OR GRAFO**
    - **KNEE IMMOBILIZER**
    - **PHYSICAL THERAPY**

---

**RESULTS - CLINICAL**
- **PRE OP FLEXION CONTRACTURE**
  - 24.9 DEGREES
- **POST OP FLEXION CONTRACTURE**
  - 5.9 DEGREES
  - **P=0.001**

---

**WALKING VELOCITY**
- **PRE OP**
  - 72.2 %
- **POST OP**
  - 80.0 %
PRE/POST GA

**MYELOMENINGOCELE**
- Gradual development
- Muscle weakness
- Increase height/weight
- Over the age of 10
- Contracture of the posterior knee capsule
  - Most important

**CONCLUSIONS**
- Posterior knee capsulectomy
- Selective lengthening of the hamstrings
- Lead to a significant improvement of the child's gait

**INDICATION FOR SURGERY**
- Sacral level
- Low lumbar level
- KFC higher than 15 to 20 degrees
- Knee flexion at mid stance > 30 degrees
- Pre op GA is important in the decision

**IF GAIT ANALYSIS IS NOT AVAILABLE**
- The degree of knee flexion during gait is double from what is measured on clinical exam
- Surgical treatment is indicated when knee flexion contracture is 15 degrees or higher
CROUCH GAIT TREATMENT

LOW LUMBAR/ SACRAL
- Knee Flexor Lengthening - Hamstring Lengthening and Posterior Capsulotomy (low level)
- Hip Flexor Lengthening (if present)

SUPRACONDYLAR EXTENSION
- INTERNAL FIXATION RIGHT ANGLE PLATE
- NO NEED FOR PATELLA ADVANCEMENT
- LLC 6 WEEKS
- GROUND REACTION AFO

CASE 5: HIP AND KNEE FLEXION CONTRACTURE CROUCH GAIT
- Age: 13 years
- High sacral level
- Bilateral hip flexion contractures
- Bilateral knee flexion contractures
- FMS 5/5/3
- No Shunt

CASE 5: PRE-OP ROM/MMT

CASE 5: PRE-OP KINEMATIC DATA
CASE 5: HIP AND KNEE FLEXION CONTRACTURE (SF)

- Surgical Treatment
  - Bilateral iliopsoas lengthening above the brim
  - Bilateral knee flexor lengthening

- Post Op
  - Total body splint-10 days
  - Long leg cast- 3 weeks
  - Knee immobilizer
  - Physical therapy strengthening
  - Gait training

CASE 5: POST-OP ROM/MMT

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<th>Range of Motion *</th>
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<td>Abduction (Hip controlled)</td>
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<td>80</td>
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<tr>
<td>Adduction (Hip extended)</td>
<td>-</td>
<td>-</td>
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<tr>
<td>Abduction (Hip flexed)</td>
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<td>-</td>
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<td>Flexion/extension</td>
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<td>Knee flexor contracture</td>
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<tr>
<td>Oblique</td>
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**STRENGTH**

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<td>Tensor</td>
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<td>30</td>
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<tr>
<td>Patellar</td>
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**TIBIAL TORSION MAY LEAD TO:**

- Significant gait abnormalities
- Biomechanics
- Velocity
- Frequent falls (internal)
- Difficulty with brace tolerance
- Skin breakdown & excessive shoe wear
- Pain: knee/foot
- Hindfoot valgus → pes planus
- Lever arm disease

**CONTROVERSIES IN ORTHOPAEDIC MANAGEMENT OF PATIENTS WITH MYELOMENINGOCELE**

Rachel M. Thompson, MD

Rotational deformities

Torsional deformities of the tibia are common in patients with spina bifida

- Internal tibial torsion
  - Fixed, congenital deformity
  - Frequently associated with clubfoot

- External tibial torsion
  - Acquired deformity likely due to muscle imbalance
  - Often associated with shortened fibula and valgus deformity of the ankle

* Neither will spontaneously resolve
TREATMENT FOR AMBULATORY PATIENTS

- **Goals:**
  - minimize bracing requirements
  - achieve as normal a gait pattern as possible

- **Surgical consideration when gait function is impacted**
  - Internal → frequent tripping 2/2 difficulty with foot clearance
  - External → crouch gait (lever arm disease)
  - External → knee valgus stress

---

**LEVER ARM DISEASE**

- **ETT > 20°** may lead to crouch
  - AFO unable to achieve extension moment for knee during stance

- **Improving ETT will improve effectiveness of AFO in achieving knee extension**

- **Internal rotation osteotomy to allow for knee extension during stance phase**

---

**VALGUS KNEE STRESS**

- Excessive ETT can also lead to valgus stress at the knee joint
  - Increased ETT likely to result in abnormal internal varus knee moment
  - TFA > 20° significantly increases this stress
  - *not controllable with orthotics over 20°*

- Abnormal stress may predispose the knee to late arthrosis

---

**VALGUS KNEE STRESS**

- **Instability, pain, arthritis in adulthood**
  - 72 community ambulators > 23 years old
  - 24% reported significant knee symptoms

- **WITH weakness of hip abductors, gastroc-soleus → characteristic gait pattern:**
  - Abductor lurch
  - Knee in valgus + flexion during stance
  - To propel forward: swivel push-off on fixed pronated foot

---

**Valgus Knee Stress**

Gait analysis: improved understanding
Identification of multiple factors → valgus stress

---

**VALGUS KNEE STRESS: TREATMENT**

- **Correct rotational deformities**
  - Leads to significant improvement in knee stress and pain
  - May prevent late degenerative changes

- **Must also correct:**
  - Knee Flexion Contracture
  - Hindfoot Valgus

- **Encourage use of AFO + crutches**
  - Pelvic obliquity/rotation
  - Stance-phase stability
  - Stress at knee joint

---
NON-OPERATIVE TREATMENT
Twister cables attached to AFOs
- Controls in/out-toeing
- Improve gait
- Not expected to correct underlying osseous deformity
- Use until patient old enough for surgical correction

OPERATIVE TREATMENT
- Indications:
  - > 6 years old
  - Severe rotational deformity (usually >20°)
  - Gait abnormalities
  - Difficulty with orthotic fitting/ skin breakdown
  - Pain
- 3D gait analysis pre-operatively: document gait pattern and help determine extent of correction needed

TIBIA DERO TATIONAL OSTEOTOMY
Distal vs. Proximal
- Krengel & Staheli: improved complication profile with distal osteotomy
- Selber: major complication rate distal 5.3%
- Mednick/Swaroop/Dias: major complication rate distal 3.1%

Plate vs. K-wire fixation
- Ryan et al: delayed union rate with k-wire fixation 4%
- Mednick/Swaroop/Dias: delayed union rate with plate fixation 2.3%
- Martin et al: complication rate with k-wire fixation 33%
- Mednick/Swaroop/Dias: total complication rate with plate fixation 33%

Tibia Derotational Osteotomy
Distal vs. Proximal
Plate vs. K-wire fixation
- Tibia only vs. Tibia & fibula osteotomy
  - Manouel & Johnson, Raffey & Hyndman: Higher trend toward posterior/coronal angulation at osteotomy with fibular osteotomy
  - Mednick/Swaroop/Dias: malunion 0%

OPERATIVE TECHNIQUE
- Distal tibia and fibula derotation osteotomy
- Tibia osteotomy just above the distal tibial physis
- Fibula osteotomy through a separate incision
- Osteotomy created using multiple drill holes
- AO limited-contact dynamic compression plate (5- or 6-hole plate)
- Incision closed over a drain with interrupted, non-absorbable sutures
**POST-OPERATIVE CARE**

- Short leg cast
  - First 3 weeks: NWB
  - Second 3 weeks: WBAT
- Cast and sutures removed at ~6 weeks
- Placed into AFO
- Aggressive rehab

**ASSOCIATED DEFORMITIES**

- Important to recognize any associated osseous deformity or muscle imbalance:
  - ITT: spastic anterior tibialis may require tenotomy
  - ETT: look for any concomitant hindfoot valgus

**FUNCTIONAL RESULTS**

- Surgical correction of torsional deformities →
  - Improved gait parameters
  - Improved range of motion
  - KAFO → AFO requirement post-operatively
- ETT:
  - Osteotomy improves knee extension during stance
  - May delay/prevent late degenerative changes in knee

**OUTCOMES OF TIBIAL DEROATIONAL OSTEOTOMIES PERFORMED IN PATIENTS WITH MYELODYSLASIA**

Rachel E. Mednick, MD, Erik B. Eller, MD, Vinoeta T. Swaroop, MD, and Luciano Díaz, MD

- Retrospective chart review
- All patients with neural tube disorders undergoing distal tibia derotation osteotomy between 1985 – 2010
  - Lumbar or sacral level myelodysplasia
  - Symptomatic deformity: >20°
  - Affecting brace wear, gait biomechanics or velocity
  - >5y age
  - >2y follow-up

**RESULTS**

82 patients (129 limbs)

- Average age at index surgery: 9.95 years
- Average follow-up from index surgery: 7.15 years

- Of the 128 limbs
  - 29% correction of internal tibial torsion
  - 71% correction of external tibial torsion
  - Average derotation: 27.5° (+/- 12.3°)
- Results maintained at final f/u: 24.5° (+/- 13°)

**Despite functional improvement...**

**Reported complication rates widely variable:**

- Mixed idiopathic/neuromuscular population: 4.8-13%
  - Myelodysplasia population: 28-33%
  + 31% re-operation rate
All superficial infections treated successfully with oral antibiotics. No effect on complication rate (p = 0.37).

Swaroop Rattey


REFERENCES


RESULTS: COMPLICATIONS

Surgical Complications

<table>
<thead>
<tr>
<th>Number (%)</th>
<th>Number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total Complications</td>
<td>53</td>
</tr>
<tr>
<td>Major</td>
<td>33.89</td>
</tr>
<tr>
<td>Fracture</td>
<td>2</td>
</tr>
<tr>
<td>Deep Infection</td>
<td>3</td>
</tr>
<tr>
<td>Hardware Failure</td>
<td>1</td>
</tr>
<tr>
<td>Minor</td>
<td>1.58</td>
</tr>
<tr>
<td>Superficial Infection</td>
<td>3</td>
</tr>
<tr>
<td>Wound Dehiscence</td>
<td>1</td>
</tr>
<tr>
<td>Pseudocyst</td>
<td>2</td>
</tr>
<tr>
<td>Delayed Union</td>
<td>3</td>
</tr>
</tbody>
</table>

• All superficial infections treated successfully with oral antibiotics
• Deep infection required surgical debridement without ROH
• 1 fracture occurred about the plate 4 months post-surgery, treated with callus distractor

RESULTS: RE-DEROTATION

<table>
<thead>
<tr>
<th>Number (%)</th>
<th>Number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total Number of Re-derotations</td>
<td>21</td>
</tr>
<tr>
<td>Internal Tension</td>
<td>21</td>
</tr>
<tr>
<td>Internal Tension</td>
<td>0</td>
</tr>
<tr>
<td>Lumbar Level</td>
<td>14</td>
</tr>
<tr>
<td>Sacral Level</td>
<td>7</td>
</tr>
<tr>
<td>Age (in years)</td>
<td>13.6</td>
</tr>
</tbody>
</table>

Repeat derotation surgery occurred at 5.0 ± 2.5 years after index surgery.

Total reoperation (complications & re-derotation) = 20%

REFERENCES


CONTROVERSIES IN ORTHOPAEDIC MANAGEMENT OF PATIENTS WITH MYELOMENINGOCELE

Foot deformities
Vineeta T. Swaroop, MD
FOOT DEFORMITY

- Foot deformity is present in almost all patients with MM
  - Can interfere with bracing to allow ambulation
  - Cause difficulty with shoe wear
  - Create cosmetic problems
  - Lead to pressure sores

FOOT DEFORMITIES

- Goals of treatment:
  - Plantigrade, flexible, braceable foot
  - Preserve range of motion
  - Avoid pressure sores

FOOT DEFORMITIES

- Basic principles:
  - Intervene early to maintain flexible foot/ prevent fixed bony deformity
    - Serial casting
    - Bracing
    - Surgery
  - Tendon excisions
  - Osteotomies allow correction while preserving joint motion
  - AVOID ARTHRODESIS
  - Use AFO after surgery to maintain correction

FOOT DEFORMITIES

- Basic principles:
  - AVOID ARTHRODESIS
    - Resulting stiffness + insensate foot
      - Neuropathic skin changes
      - Pressure sores
      - Osteomyelitis
      - Amputation
    - When needed, bony procedures should be extra-articular, preserving motion

CLUBFOOT

- Most common foot deformity in MM
  - 30-50% of patients with MM

- Incidence varies with neurologic level
  - 90% thoracic/lumber
  - 50% sacral

CLUBFOOT

- Different from idiopathic clubfoot:
  - Severely rigid deformity
  - Recalcitrant to treatment
  - Propensity to recur
  - Often associated with severe internal tibial torsion

References:
Westcott et al Radiographics 1992
de Carvalho Neto et al J Pediatr Orthop 1996
CLUBFOOT - ETIOLOGY

- Multifactorial
  - Spasticity
  - Intrauterine positioning
  - Contractures
  - Muscle imbalance

- i.e. Low-lumbar level patient:
  - Clubfoot may develop in part due to retained activity/contracture of tibialis anterior + functional absence of peroneals

CLUBFOOT - TREATMENT

- Traditional teaching = non-surgical management rarely successful
  - Splinting
  - Serial casting
  - Stretching
  - Extensive soft tissue release (PMLR) surgery necessary for correction

CLUBFOOT - TREATMENT

Ponseti method

- Multiple studies have reported early results in clubfeet associated with MM
  - No long-term follow-up

- What is standard of care for clubfoot treatment in spina bifida?

- Should Ponseti method be attempted in all patients?

CLUBFOOT - PONSETI METHOD

Ponseti method:

- 28 clubfeet in MM
- Initial correction achieved in 27/28 (96%)
- Average follow-up 34 months:
  - Relapse occurred in 68%
  - 4/28 (14%) required extensive soft tissue releases
- Complications:
  - 9 feet (32%) blistering in foot abduction orthosis
  - 6/9 had recurrence while out of brace requiring repeat casting
  - 3 feet (11%) cast slippage leading to complex clubfoot deformity
  - 2 patients with distal tibial fractures


Ponseti method:

- 9 clubfeet in MM
- Initial correction achieved in all feet
- Average follow-up 33 months:
  - 5 feet (56%) with recurrence
  - 3 (33%) required extensive soft tissue releases
- Complications:
  - 2 feet (22%) skin breakdown

Janicki et al J Pediatr Orthop 2009

Ponseti method:

- 24 clubfeet in patients with ‘spinal dysraphism’
- Initial correction achieved in 22/24 (92%)
- Average follow-up 4 years:
  - 13/22 feet (60%) with recurrence
  - Relapse occurred at mean of 3.2 years after initial correction
- Complications:
  - Pressure sores in 4 feet

Dunkley et al J Child Orthop 2015
Ponseti method:
- Initial correction in 92-100% feet
- 56-68% recurrence
- High rate of complications: skin breakdown, fractures
- Ponseti method can be useful in decreasing need for extensive soft-tissue release
- BUT families should be counseled about:
  - High risk of recurrence
  - Potential for need for further treatment
  - Risk of skin breakdown, fractures

If soft-tissue release is indicated →
- Surgical treatment should consist of a radical posteromedial-lateral release using a cincinnati incision
- All tendons excised (not lengthened)
- Subtalar, calcaneocuboid, and talonavicular joints completely released
- Separate plantar release may be needed through plantar incision
- Optimum age is 10-12 months

Improved results have been shown with use of temporary k-wire to derotate talus in ankle mortise
- K-wire placed into posterolateral aspect of talus
  - Rotate talus medially
  - Navicular reduced on talar head
- Second k-wire driven through body of talus into navicular to hold reduction
- Temporary k-wire removed
- Use additional k-wire to maintain talocalcaneal alignment

Long leg posterior mold splint for 1st 2 weeks
- Foot in slight equinus
- Decrease tension on interrupted sutures used for skin closure
- Change to long leg cast with foot in corrected position x 6 weeks
- Day and night-time AFOs used to maintain correction for long-term

Good results reported in 61-83% of patients
- de Carvalho Neto et al J Pediatr Orthop 1996
- Flynn et al J Pediatr Orthop B 2004

Partial or complete recurrence occurs in 20-50% after primary surgical correction
- Adduction deformity common
- Treat with “double C osteotomy”
- For complete recurrence → taelectomy
CLUBFOOT SUMMARY

- What is the standard of care?

  **Ponseti**
  - Initial correction in 92-100% feet
  - 56-68% recurrence
  - High rate of complications: skin breakdown, fractures

  **PMLR**
  - Good results in 61-83% (varies by motor level)
  - Partial or complete recurrence in 20-50%

VERTICAL TALUS

- Rigid rocker-bottom flatfoot deformity

- ~10% patients with MM

- Occurs in 2 forms:
  - Congenital – more common
  - Developmental

PATHOANATOMY

- Fixed hindfoot equinus, valgus
  - Due to contracture of Achilles and peroneals

- Rigid midfoot dorsiflexion
  - Due to dislocation of navicular

- Forefoot abducted, dorsiflexed
  - Due to contractures of tibialis anterior and toe extensors

CLINICAL EXAM

- “A prominence in the sole from which the heel and forefoot rise in a gentle curve.”
  - Lloyd-Roberts and Spence

- ‘Persian slipper’

- Few posterior heel skin creases
- Head of talus palpable on plantar medial aspect of foot
  - Convex plantar surface
  - Crease overlying sinus tarsi

- Not correctable by manipulation!
  - Cannot create longitudinal arch
  - Cannot reduce head of talus
  - Cannot dorsiflex ankle
CLINICAL EXAM

- Rigid rockerbottom foot
- Hindfoot: equinus, valgus
  - Achilles tendon contracted
- Midfoot: abducted, dorsiflexed
- Forefoot: abducted, dorsiflexed

RADIOGRAPHIC EXAM

- AP, lateral
  - Diagnosis confirmed with:
    - Lateral in forced plantar flexion
    - Lateral in forced dorsiflexion
- AP:
  - Midfoot valgus
  - Talocalcaneal angle > 40°
    (normal = 20-40°)

RADIOGRAPHIC EXAM

- Before ossification of navicular, 1st MT used as proxy
- Forced plantar flexion lateral
  - Persistent dorsal dislocation of TN joint
  - Meary’s angle > 20°
- Forced dorsiflexion lateral
  - Persistence of plantar flexion of talus
  - Equinus position of calcaneus

TREATMENT

- Goal = restore normal relationship between talus, navicular, and calcaneus
  - Provide plantigrade weight-bearing surface
- Traditional treatment =
  - Complete posteromedial-lateral and dorsal release
    - Between 10-12 months of age

TREATMENT

- Dobbs method
- Serial manipulation
- Cast immobilization
- Open talonavicular pin fixation
- Percutaneous Achilles tenotomy
- Excellent short-term results

Foot stretched into plantar flexion and inversion while counterpressure applied to medial aspect of head of the talus
TREATMENT

Foot position before TN pinning:
- Maximum hindfoot varus
- Forefoot adduction
- Maximum plantar flexion

RESULTS – DOBBS METHOD (ALL ETIOLOGIES)

- 19 feet
- At least 2y follow-up
- Initial correction achieved in all cases
  - Average 5 casts
- Final f/u: mean DF 25°, mean PF 33°
- Recurrence in 3 patients
  - None had pin fixation of TN joint
- Significant improvement in all radiographic parameters compared with pretreatment
- All measured angles were within normal values for patient’s age
VERTICAL TALUS: TREATMENT

Extensive soft-tissue release:

- Single-stage surgical correction
  - Kodros, Dias J Pediatr Orthop 1999

- Preferred over two-stage procedure
  - Associated with complications: AVN of talus

RESULTS – DOBBS METHOD FOR NON-IDIOPATHIC FEET

- 25 feet (6 with MM)
- >2y follow-up after surgery
- Initial correction achieved in all cases
  - Average 5 casts
- Significant improvement in all radiographic parameters
- 5 (20%) feet had recurrence
  - No recurrence in MM feet

SINGLE-STAGE CORRECTION

- Cincinnati incision
- Achilles z-lengthened
- Posterior capsules of tibiotalar and subtalar joints opened
- Circumferential release subtalar joint
- Release talonavicular joint
  - Medial/dorsal
- Release calcaneocuboid joint if needed
- Anterior (and posterior if needed) tibial tendons detached/transfered

SINGLE-STAGE CORRECTION

- K-wire placed into posterolateral talus
- Joystick to elevate talus
- While plantarflexing navicular and forefoot
- Talonavicular and subtalar joints pinned
- Extensor, peroneal tendons lengthened as needed thru 2nd incision

SINGLE-STAGE CORRECTION

- Very similar to clubfoot surgery…
- Main difference is rotation of the talus:
  - Dorsiflexion instead of internal rotation
SINGLE-STAGE CORRECTION

- Maintain elevation – prevent swelling
- Cast change in 2 weeks

SINGLE-STAGE: RESULTS

Kodros, Dias J Pediatr Orthop 1999
- 42 feet
- 100%: good or fair results at final f/u
- No wound complications or avascular necrosis of talus
- Mild pain in 3 feet
- All patients/ families satisfied with results

REFERENCES