Long-Term Management of Spina Bifida Patients

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Life Expectancy

54% reached 19th year
A study of deaths and handicap in a consecutive series of spina bifida/teakle... from birth. Z Kinderchir 38 Suppl 2-103-2, 1983
56/117 died 22-28 yrs after closure. 33/61 of survivors were living independently. Two main determinants for functional independence were IQ and neurological deficit.

Nature of Disability

Long-term data is limited

Neurologic
tethered spinal cord, hydrocephalus
Orthopedic
scoliosis, spinal instability
Urologic
Functional
employment
social
access to health care

Wheelchair dependent 35.9%
Shunt present 36.7
Scoliosis 47
Joint deformities & contractures 66.8
Intermittent catheterization 28
Urinary diversion 21.8
Abnormal renal function 46.1
Hypertension 14.5
Epilepsy 8.8

Life Satisfaction

179 young adults with spina bifida (79% SB aperta, 67% hydrocephalus, 39% wheelchair bound)
24% were satisfied with their lives
emotional health scores did not differ
Tethered Spinal Cord (TSC)

Clinical Presentation Varies by Age

Infants
- cutaneous stigmata, urinary disturbance, lower extremity deformity

Young children
- motor and sensory symptoms
- gait disturbance, progressive LE weakness
- regression in bladder control
- sensory loss in a non-segmental distribution

Tethered Spinal Cord (TSC)

Clinical Presentation Varies by Age

Teenagers
- pain, urinary dysfunction, scoliosis
  - pain is in lower back, perineum and non-dermatomal

Adults
- pain is a primary presenting symptom
- urinary dysfunction very common

Indications for Surgery

Separate tethered cord from spinal deformity

Tethered spinal cord
- progressive neurologic deficit
- pain (radicular vs. mechanical)

Spinal deformity
- progressive scoliosis (>30 degrees)
- instability
- severe mechanical back pain

Neurologic Deficit

Most important factor is impact upon functional capacity

Sensory change or limited change in motor function should not drive decision-making

Cohort of 10 patients
- curvature improved in all; more caregiver assistance required in first 6 months; 12 months to recovery
- ambulation more difficult in those walking prior to surgery
- 80% complication rate


Tethered Cord Release & Scoliosis

Progression of scoliosis plateaued or declined following release of tethered cord in patients with lumbar and sacral level lesions.

Release did not halt the progression of scoliosis in the thoracic level group.

Tethered cord release altered the course of lordosis in L1 through L3 level lesions, but did not affect progression of lordosis in patients with L4, L5 or sacral level lesions.

Reigel DH. Change in spinal curvature following release of tethered spinal cord associated with spina bifida. Pediatric Neurosurgery 20:34, 1994
Spinal Deformity Correction

Untethering should be considered prior to deformity correction or spinal fusion; particularly in the following situations:

1. Co-existing neurological symptoms or pain
2. Intent to correct coronal plane deformity includes plan to lengthen spine
3. Hyperlordosis or spondylolisthesis is directly in area of spinal cord tethering

Pre-Operative Evaluation

Imaging
- plain films
- MRI and CT (consider CT brain)

Urodynamics

Functional analysis
- ambulation - gait analysis (evaluation by physiatrist)
- self-care
- pre-op consultation with pain service

Pre-Operative Evaluation

Treatment - Pain
- Neurogenic pain: Neurontin, desensitization therapy, TENS
- Mechanical pain: Steroids, anti-inflammatory agents

Treatment - Functional improvement
- Physical therapy
- Psychotherapy

Operative Treatment

Goal - Remove tension and compression without additional spinal cord trauma

1. Identification of normal spinal cord and nerve roots
2. Separation of scar from distal cord segment
3. Debulking of lipoma
4. Reconstruction of dura (consider Gore-tex patch)

Intra-op considerations
- neuromonitoring
- prevention of CSF leak

Outcome

Presenting Symptoms/Signs  Post-op Outcome

Presentation during Adulthood

61 adults; 41% lipomyelomeningocele, 36% fatty filum terminale

Back pain improved in 65%
LE pain improved in 53%
LE weakness improved in 47%


Case 1
20 y/o old female with spina bifida and hydrocephalus with progressively increasing pain in lower extremities and limitation in ambulation - increasing time spent in wheelchair.

Case 2
34 y/o woman with spina bifida and hydrocephalus. Wheelchair dependent, worsening back pain and difficulty sitting for long periods of time.
Conclusions
Conservative management is preferred
Treatment decisions should be designed to maximize functional capacity
Realistic expectations are crucial
Multidisciplinary team; or a group of interested providers should manage these patients

References

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