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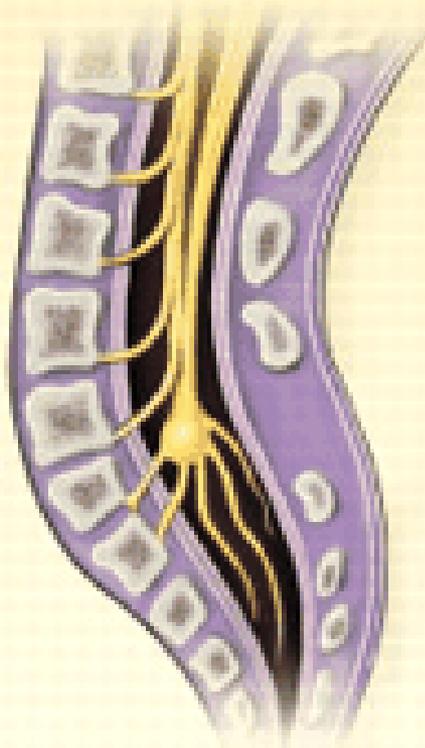
# SPINA BIFIDA: NEUROSURGICAL ISSUES AND LIFE OUTCOMES

# Introduction

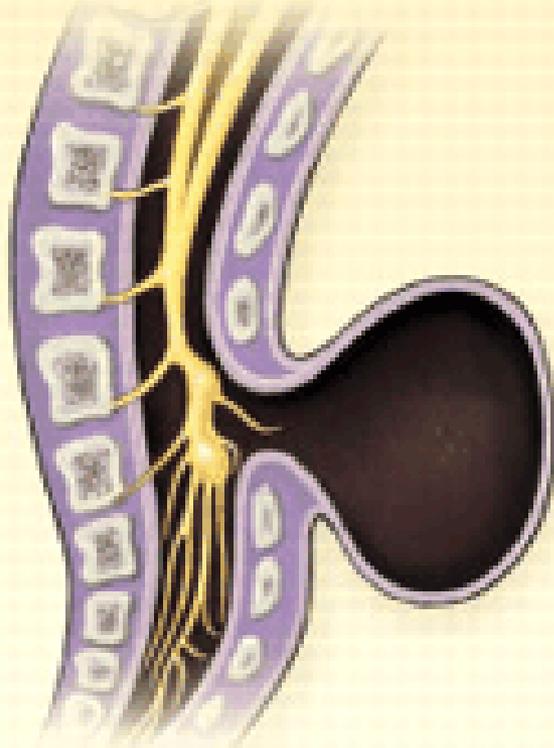
- Description of Myelomeningocele and associated neuroanatomical malformations
- Discussion of quality of life in pediatric and adult populations

# Introduction

- Recent estimates indicate that 1500 children are born with myelomeningocele (MMC) in the United States each year
- With the introduction of immediate spinal closure and shunting for hydrocephalus in newborns with MMC in the 1960s, the survival rate increased dramatically from approximately 10% to approximately 75% of individuals with MMC surviving into adulthood today
- The 1-year survival rate of infants born with MMC now exceeds 92%



**Spina bifida occulta**



**Meningocele**



**Myelomeningocele**

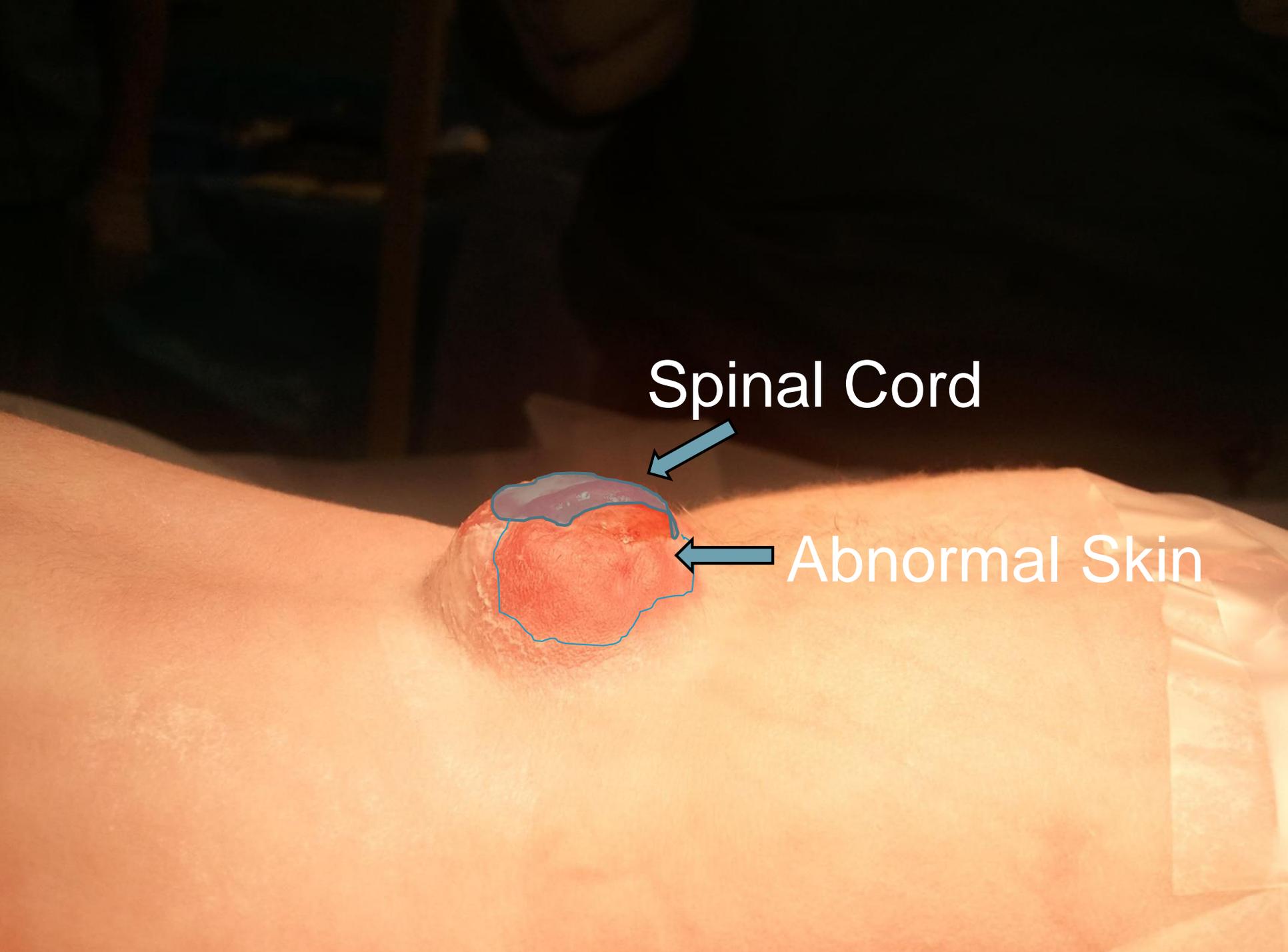


Neural Placode  
(spinal cord)



Abnormal Skin





Spinal Cord

Abnormal Skin

# MMC level

Thoracolumbar junction	45%
Lumbar	20%
Lumbosacral	20%
Sacral	10%
Other (high) lesions	5%

**Table 1. Effects of Meningomyelocele on Body Function and Structure as Well as Activities With Providers Traditionally Associated With Those Categories**

Body Function/Structure and <i>Activity</i>	Traditional Provider
Spinal Cord	
Primary abnormalities of the cord	Neurosurgeon
Split cord/Syrinx/Spinal cord atrophy/Tethering	Physical Therapist
Spine	Orthopedist
Scoliosis/Kyphosis	Orthotist
Motor nerves	
Neurogenic bowel	Nurse
Constipation	Pediatrician*
Continence/Toileting	Gastroenterologist/Surgeon
Male sexual dysfunction	Urologist
Weakness and Paralysis	Orthopaedist
Joint Contractures	Physical Therapist
<i>Mobility</i>	Orthotist
<i>Transfers/Activities of Daily Living</i>	Occupational Therapist
Neurogenic Bladder	Nurse
Urinary Tract Infections	Urologist
Genitourinary Reflux	Nephrologist
<i>Continence/Toileting/Hypertension/Renal failure</i>	Pediatrician*
Fine Motor Coordination	
<i>Activities of Daily Living</i>	Occupational Therapist
Sensory nerves	Nurse
Decubitus ulcers	Plastic Surgeon
Impaired orgasmic sex	Gynecologist
Brain	
Hydrocephalus/Shunt complications/Chiari II malformation	Neurosurgeon
Strabismus	Ophthalmologist
<i>Learning</i>	
<i>Learning Disabilities/Impaired Executive Function/Attention deficit hyperactivity disorder (ADHD)</i>	Pediatrician*
Hypoventilation	Psychologist
Sleep disorder	Pediatrician*
Impaired language skills— <i>Communication</i>	Pulmonologist/Otolaryngologist
Epilepsy	Speech and Language Therapist
Precocious puberty	Neurologist
Socioeconomic impact	Pediatrician*/Endocrinologist
Stress on families/Cost of care	Nurse
Latex allergy	Social Worker/Financial Consultant
Obesity	Nurse/Pediatrician*
<i>Social skills</i>	Nurse/Pediatrician*/Dietician
<i>Impaired social interactions</i>	Speech and Language Therapist
Wellness	Psychologist/Social Worker
Physical fitness	Nurse
Nutrition	Pediatrician*/Physical Therapist
	Dietician

Activities are shown in italics.

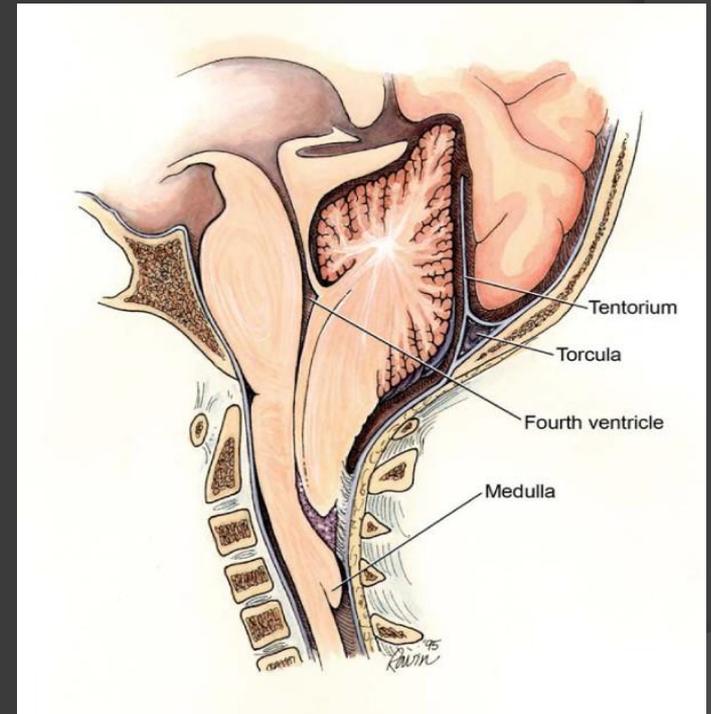
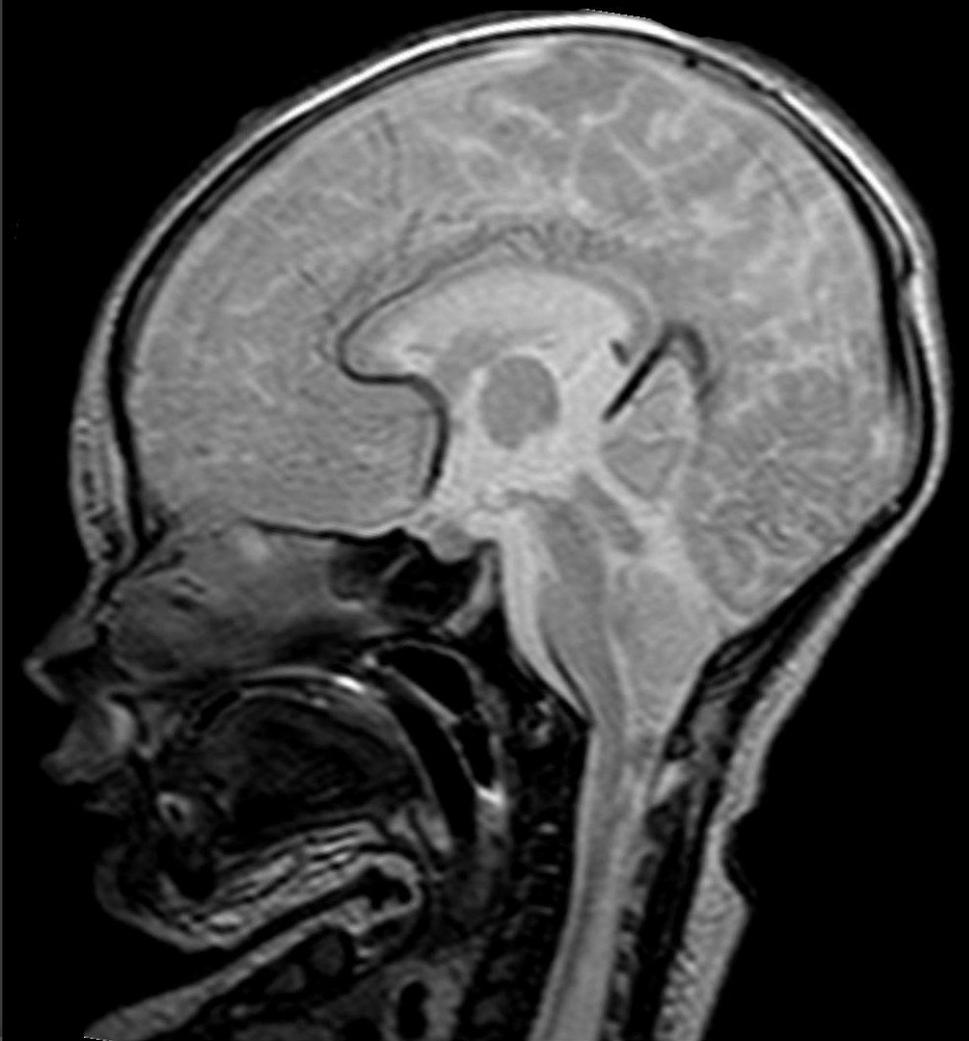
\*Includes Developmental Pediatrician or Pediatric Physiatrist.

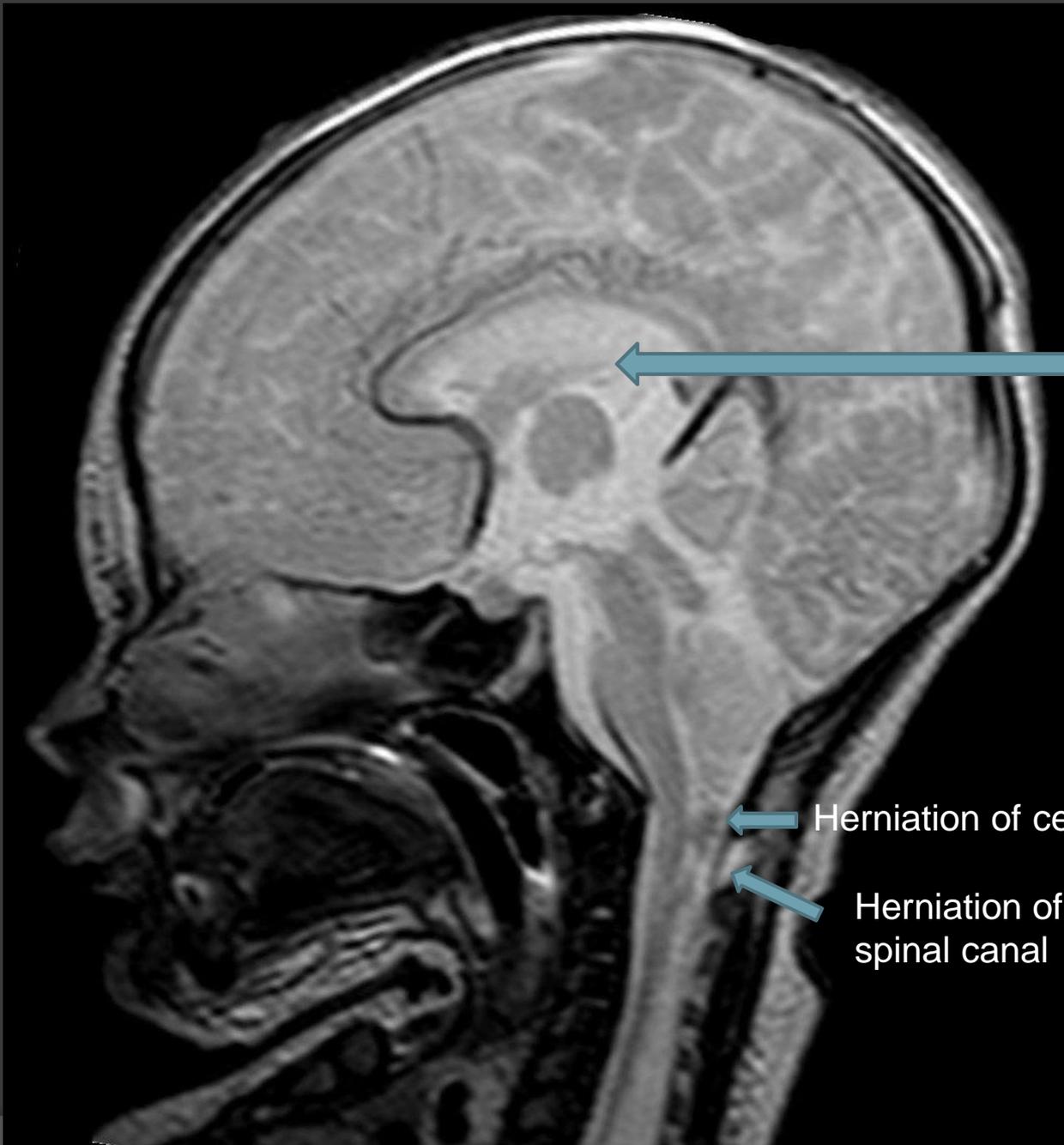
# Hydrocephalus

- Ventricular shunting of hydrocephalus occurs in approximately 85% of patients with MMC



# Chiari II Malformation





Hydrocephalus  
(enlarged ventricles)

Herniation of cerebellar vermis into spinal canal

Herniation of brainstem and 4<sup>th</sup> ventricle into spinal canal

# Chiari II Malformation

## ⦿ Signs and symptoms

- Occur characteristically within age groups

- Newborns

- Usually asymptomatic

- Infants

- Signs of brainstem compression

- Older children/young adults

- Spinal and cerebellar signs



Stridor (vocal cord paralysis)

Central sleep apnea/breath holding

Aspiration w/ dysphagia

**Nasogastric regurgitation**

Failure to thrive

Hypotonia/quadraparesis

**opisthotonus**

# Chiari II Malformation

## ⦿ Signs and symptoms

- Occur characteristically within age groups
  - Newborns
    - Usually asymptomatic
  - Infants
    - Signs of brainstem compression
  - Older children/young adults
    - Spinal and cerebellar signs



Occipital/cervical pain  
Hand weakness  
Myelopathy  
Ataxia  
Scoliosis  
dysarthria

# Tethered Cord

- ⦿ Patients with symptomatic TCS present with back pain, worsening motor weakness, or new bowel or bladder symptoms.
- ⦿ Pain is the most common presenting symptom
  - Back pain occurs in 73% of patients presenting with TCS
  - leg pain or sciatica is the presenting symptom in 56% of patients.
- ⦿ Muscular weakness and bladder dysfunction occur as the presenting symptom in 78% and 71% of patients, respectively

# Tethered Cord

- Spinal cord re-tethering occurs in approximately 23% of patients between 1 month and 23 years after surgery.



# Quality of Life (QOL)

- ⦿ Advances in medical care have improved medical outcomes in patients with MMC
  - General health
  - Longevity
- ⦿ How do people with MMC do from a quality of life standpoint?

# Pediatric QOL

- Populations of children with MMC have been studied with regard to QOL
- The following study looked at 125 pediatric patients with MMC:

## **Assessing health-related quality of life in children with spina bifida**

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# Pediatric QOL

- 86% had CSF shunts in place
- 11% had undergone Chiari decompression
- 59% had undergone at least 1 shunt revision (range 0–15)
- 47% were able to ambulate in the community.
- 23% reported bowel continence
- 29% reported bladder continence.
- 30% had undergone a tethered cord release

# Pediatric QOL

- ① The most consistent variable associated with difficulty attaining adult milestones was hydrocephalus, the presence of which reduced the likelihood of living independently, but did not contribute to reduced life satisfaction

# Pediatric QOL

- Overall QOL score was significantly associated with both bowel continence and bladder continence
- Patients with a shunt had lower overall QOL scores compared with patients without a history of shunting.
- Scores for ambulation and cognition were also lower in shunt-treated patients

# Pediatric QOL

- The presence of a shunt is simply a proxy for greater degree of illness or chronic disease

# Quality of Life (QOL)

- Populations of adult with MMC have also been studied with regard to QOL and life satisfaction.
- The following group studied 90 adults ranging in age from 25 to 85 years old:

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## **Outcome and Life Satisfaction of Adults with Myelomeningocele**

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# Adult QOL

- The presence of hydrocephalus, which occurs in approximately 80% of individuals with MMC, significantly increases the likelihood of cognitive impairment and is the most commonly reported variable associated with poor outcome

# Adult QOL

- The majority of participants had lumbo/sacral lesions and required a shunt for the treatment of hydrocephalus
- Nearly half of all participants required a wheelchair for mobility
- Most participants did not have complete bowel and bladder continence

# Education

- ◎ 94% completed high school or a high school equivalent (GED).
- ◎ 62% completed their high school education in a typical classroom setting
  - 13% required an aide or extra one-on-one assistance
  - 10% required special education for specific subjects
  - 14% were primarily in a special education classroom.
- ◎ Participants with hydrocephalus were less likely to be in a typical classroom setting in high school

# Education

- 56% completed a technical, associates or college degree.
- Participants in a typical classroom setting in high school were more likely to obtain post-secondary education
- There were no significant associations between hydrocephalus, lesion level, mobility, gender, age or continence and obtaining post-secondary education.

# Employment

- ⦿ Participants primarily received income from disability benefits (59%) and/or their own employment (48%).
- ⦿ 85% held a job at some point during their life
- ⦿ 51% worked part-time (less than 40 hours per week).
- ⦿ The most common reported reasons for part-time employment:
  - health issues 42%
  - job availability 35%
- ⦿ There was also no association between employment and gender, age, mobility, lesion level, continence, or hydrocephalus.
- ⦿ A total of 17% of participants never held a job.

# Relationships

- 43% of participants lived with their parents
- 21% lived alone with or without assistance.
- Individuals who were independently mobile and those without hydrocephalus were more likely to live independently
- There was no association between participants' living situations and continence, lesion level, age or gender.

# Relationships

- Over half (52%) of participants never had a serious partner or spouse, while 28% had been married.
- While the rate of marriage was low, for those who did marry, the age of marriage was consistent with the general population.
- Seven participants (17%) reported having a partner with a disability including NTDs, blindness and cerebral palsy.
- Individuals without hydrocephalus and with independent mobility were more likely to be in a relationship. There was no association between relationship status and age, gender, lesion level or continence

# Reproduction

- ⦿ Though the majority of women with MMC do not have children, there is no evidence suggesting decreased fertility in this population.
- ⦿ Women with MMC have normal menstruation, are able to become pregnant and have healthy babies
  - These women have unique concerns, including recurrent urinary tract infections, worsening kidney function, and premature labor

# Reproduction

- In contrast to women, data show that men with MMC have reduced fertility, but those with lower level lesions can successfully reproduce

# Reproduction

- Additionally, both men and women with MMC are at increased risk to have a baby with an NTD

● Questions?