



Spina bifida
Early management and
long-term care

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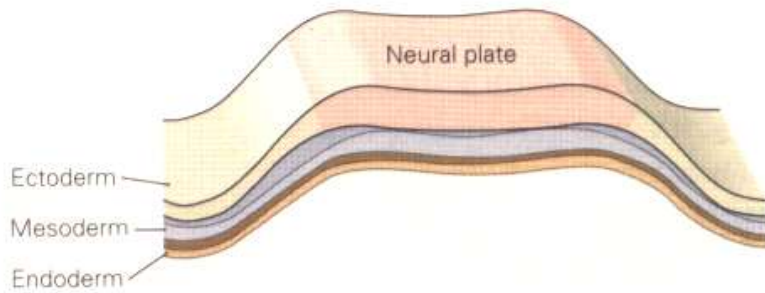
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Perspective

- Spina bifida is the most common birth defect involving the Central Nervous System
- Often *preventable*
- Initial management has a *profound* effect on survival and the handicaps that they may suffer
- Hope and support make this a manageable condition which culminates in a productive and meaningful life for many...

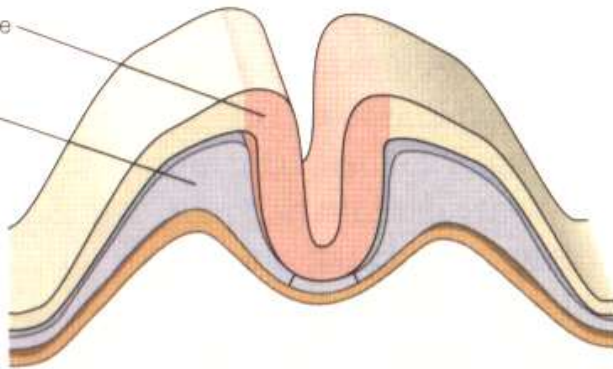
Meningomyelocele is the most complex congenital abnormality compatible with a normal life (David McLone)

A



B Neural groove

Paraxial mesoderm



Normal embryology: Closure of the neural tube

Principles of Neural Science
Kandel ER
2000

This process is complete within
the first month post-conception;

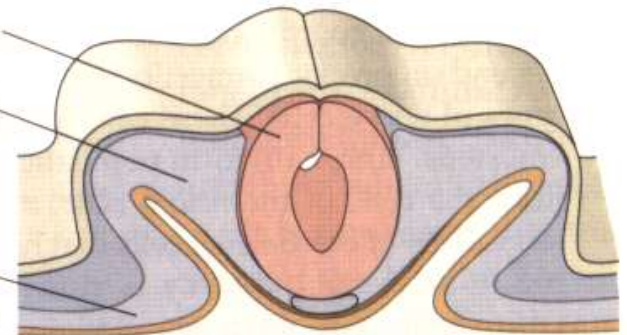
This has clear implications
for the concept of prevention
(see later)

C

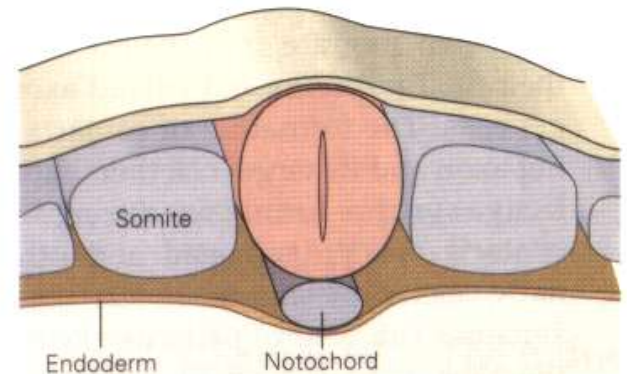
Neural tube

Paraxial mesoderm

Lateral plate mesoderm



D



This is a real process-
not just a hypothetical
construct!

Scanning
Electron
Micrograph
of
Chick
Neural
Tube



Important points to remember about neural tube closure

- Occurs by **day 28** postconception.....
this is critical for understanding strategies for prevention!
- This process is called **primary neurulation** and gives rise to virtually the entire CNS; the terminal part of the spinal cord arises through caudal regression or **secondary neurulation**
- Abnormalities cause conditions that are termed neural tube defects (NTDs) or **dysraphism** and may occur at either end

Classification of Neural Tube Defects

Cranial dysraphism

open *anencephaly*

closed *encephalocele*

Spinal dysraphism (*Spina bifida*)

open *spina bifida aperta*
 meningomyelocele (myelomeningocele)

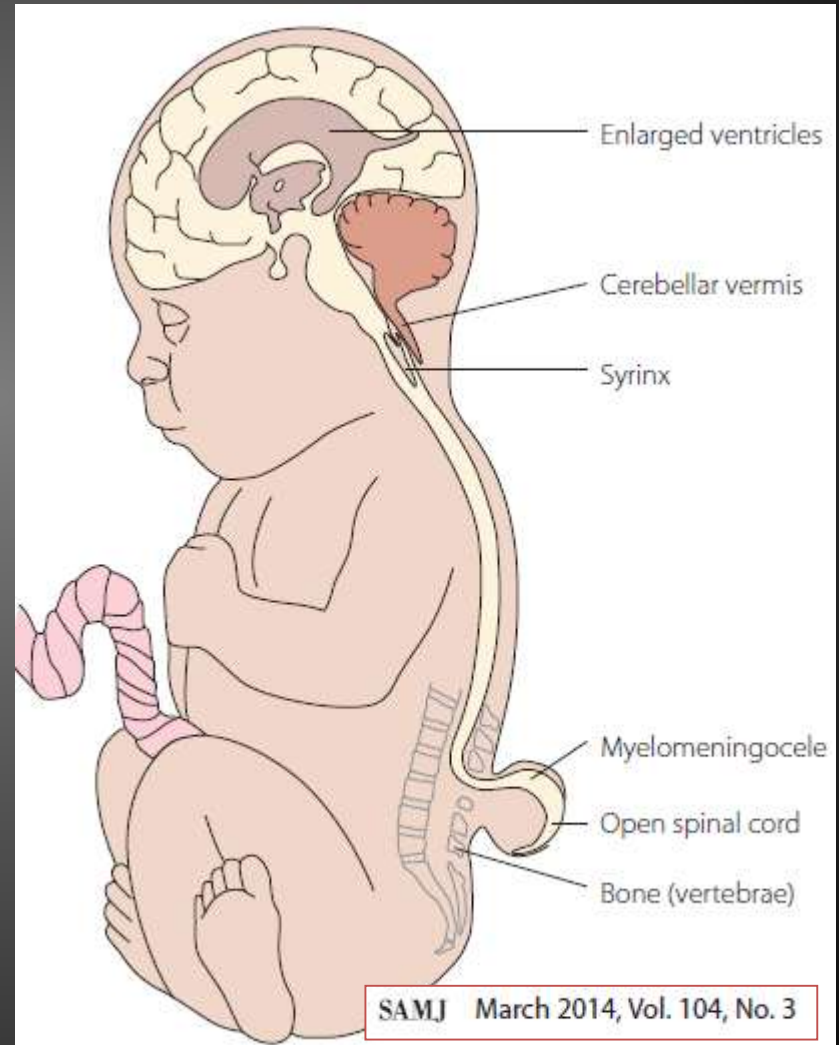
closed *spina bifida occulta*

Encephalocele

- **Anencephaly** is probably the commonest NTD, but seldom seen clinically as it usually results in a miscarriage or termination if diagnosed antenatally
- **Encephalocele** is usually obvious at birth and may be
 - **Anterior** long term prognosis usually good
 - **Posterior** long term prognosis poor in most cases due to extent of disruption of brain development; some may do well though
 - **Basal** very rare

Spinal Dysraphism

Meningomyelocele (open spina bifida)

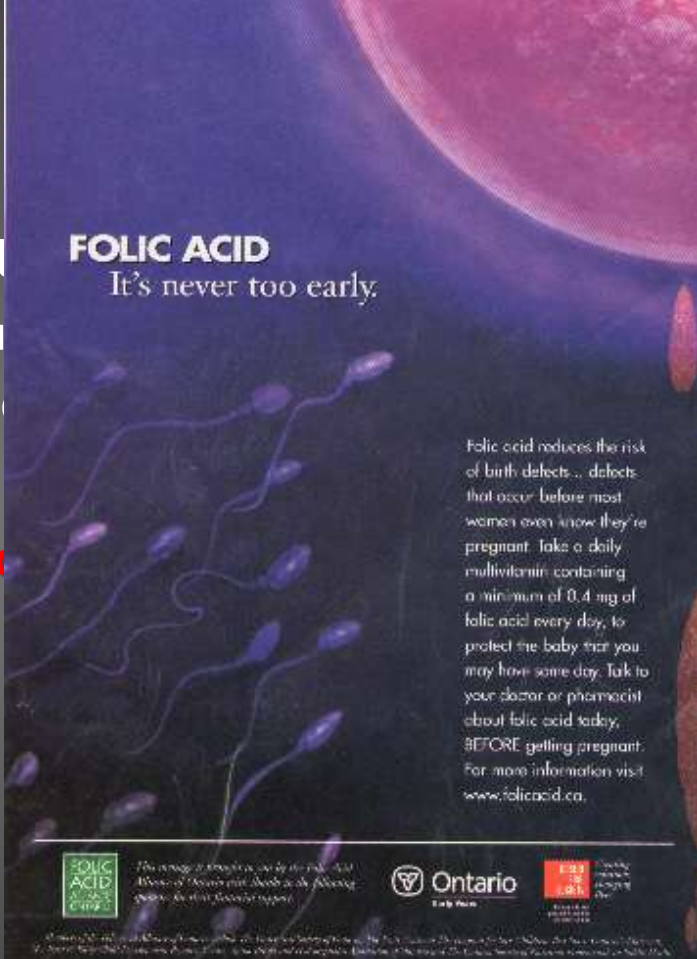


There is a **very** wide variation in size, level and appearance...



Perinatal management

- **Antenatal diagnosis**
 - NTD incidence varies the country
 - 2/1000 live births Cape Town (But)
 - family history, many other risk factors
 - Investigations
 - Screening: **maternal serum alpha-fetoprotein**
 - Diagnostic: **ultrasound**
- **Labour and delivery**
 - NVD vs Caesarean Section



FOLIC ACID
It's never too early.

Folic acid reduces the risk of birth defects... defects that occur before most women even know they're pregnant. Take a daily multivitamin containing a minimum of 0.4 mg of folic acid every day, to protect the baby that you may have some day. Talk to your doctor or pharmacist about folic acid today. **BEFORE** getting pregnant. For more information visit www.folicacid.ca.

FOLIC ACID 0.4 mg

The average woman is low in folic acid. Because of this, many women do not know they're pregnant. Take a daily multivitamin.

Ontario Early Years

ESB Early Start

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- **The folate story... *pre-conception reduces the risk by >70%!***

Decline in the Prevalence of Neural Tube Defects Following Folic Acid Fortification and Its Cost-Benefit in South Africa

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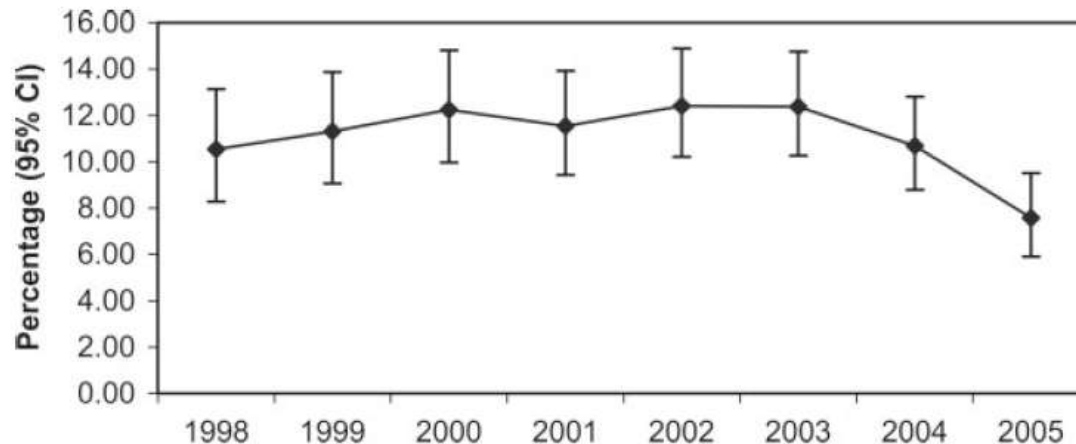


Figure 1. Proportion of NTD deaths to all other congenital anomalies, <1 year, South Africa 1998–2005.

What about 2014?

Surgical repair of meningocele

Goals:

1. freeing of the neural placode
2. neurulation
3. watertight closure of the dura
4. skin closure



Neural placode
(Open Spinal Cord)



Dura mater



and that's not all folks.....

Postoperative management

- Keep flat for a week to avoid CSF leak
- Normal feeds- ensure no bulbar palsy
- Daily head circumference (watch for hydrocephalus)
- Counsel parents about folate prophylaxis in future
- **Multidisciplinary follow-up....**
 - Neurosurgery, Orthopaedics, Urology, Radiology,*
 - Developmental Paediatrics, Genetics,*
 - Stomatherapists, Physiotherapists*
 - Community Nurses, Social Workers....*

Is non-operative management an option?

The placode may epithelialise....



If a decision is made **not** to treat a severely affected newborn, this must be thought through very carefully as the child may survive, culminating in a much worse situation with a massive sac, sepsis etc

Changing philosophies of management

- Pre-1950 survival <10%
- Shunts
- 1970s Lorber “Selective treatment”
- 1990s Groningen protocol



Casey Holter

Deliberate termination of life of newborns with spina bifida, a critical reappraisal

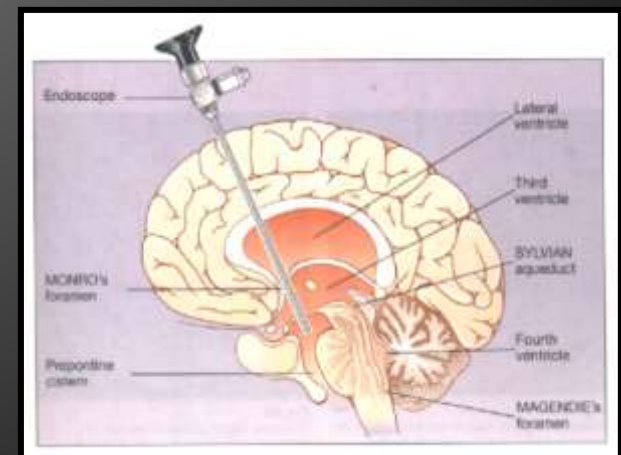
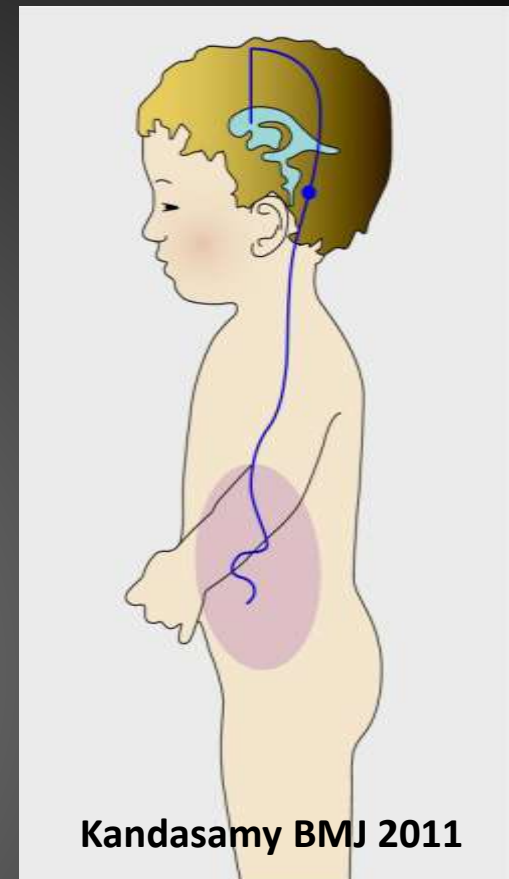
T. H. Rob de Jong

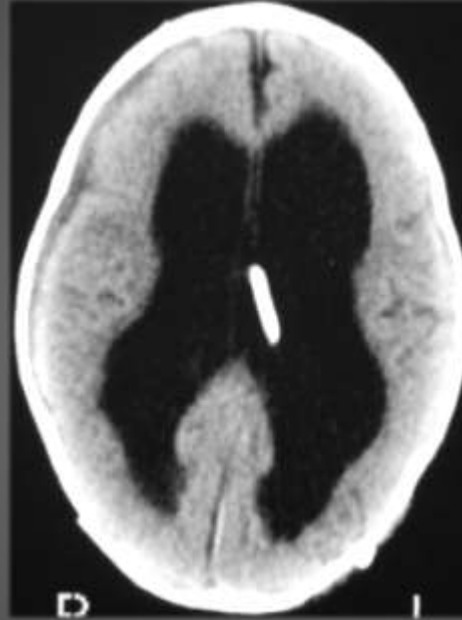
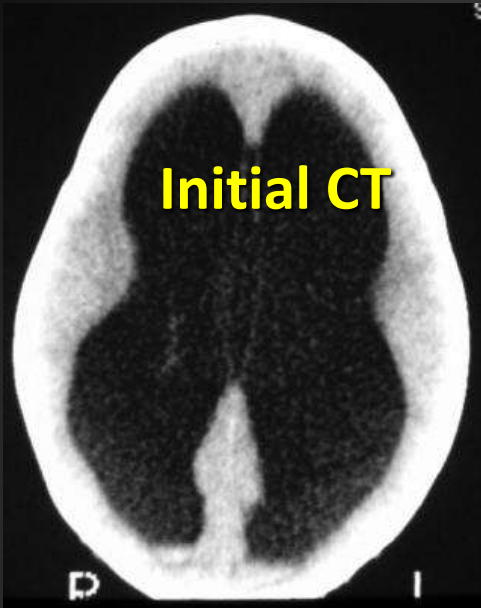
Long term issues

- Neurosurgical
 - Chiari malformation
 - Hydrocephalus
 - Tethered spinal cord
- Orthopaedic
 - Feet, ankles, knees, hips..
 - Spinal deformity
- Urological
- Cognitive

Hydrocephalus

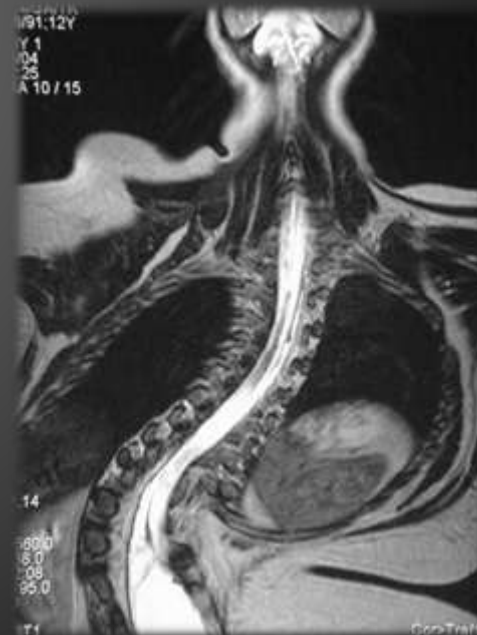
- *A hydrodynamic disorder of CSF circulation leading to an increase in intracranial pressure*
- Treatment options
 - Ventriculoperitoneal shunt
 - Endoscopic third ventriculostomy





Orthopaedic issues

- Lower extremity
 - Equinovarus (clubfoot) in >50%
 - Other foot and ankle, knee and hip deformities
- Spine
 - Incomplete posterior arch
 - May have other bony anomalies
 - Scoliosis
 - kyphosis



Urological issues

- **Neurogenic bladder dysfunction**
 - Incontinence
 - Vesico-ureteric reflux (3-5% 40-50% if not correctly managed)
- ***Proactive management***
 - urodynamics
 - anticholinergics
 - **clean intermittent catheterisation** (CIC: Lapedes 1971)
 - Vesicostomy, bladder augmentation etc
- **Don't forget**
 - Constipation
 - Sexual function

Cognitive outcome

- Mean IQ of patients with spina bifida is within the **normal range**, but usually lower than that of the general population
- Weaker in visuo-spatial and motor domains
- Discrepancy between Verbal IQ and Performance IQ
- Numerous factors play a role, including **hydrocephalus** and shunt complications, lesion level, other complications, social circumstances and expectations etc

Spennato and Cinalli 2008

Chicago series: the gold standard!

118 children managed actively from 1975-1979

19 lost to follow-up

28 died

71 reviewed at 20 – 25 years

63% normal schooling (37% special classes)

45% actively employed (others volunteers)

Pediatr Neurosurg 2001;34:114–120

Spina Bifida Outcome: A 25-Year Prospective

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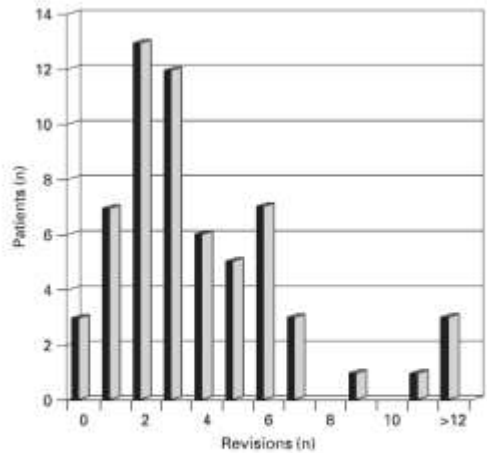


Fig. 1. The number of shunt revisions completed on the study cohort.

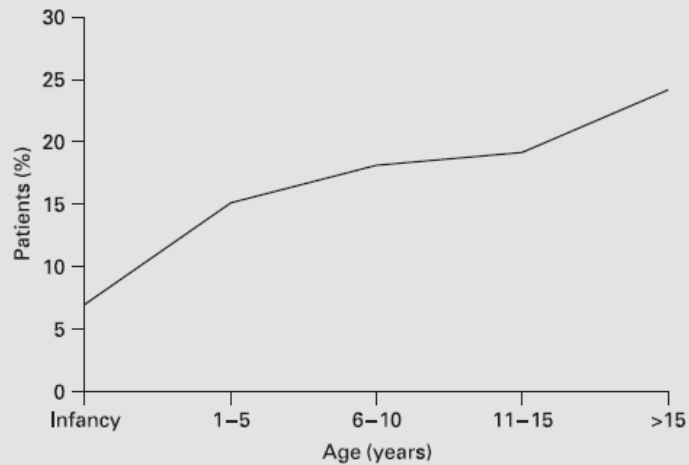
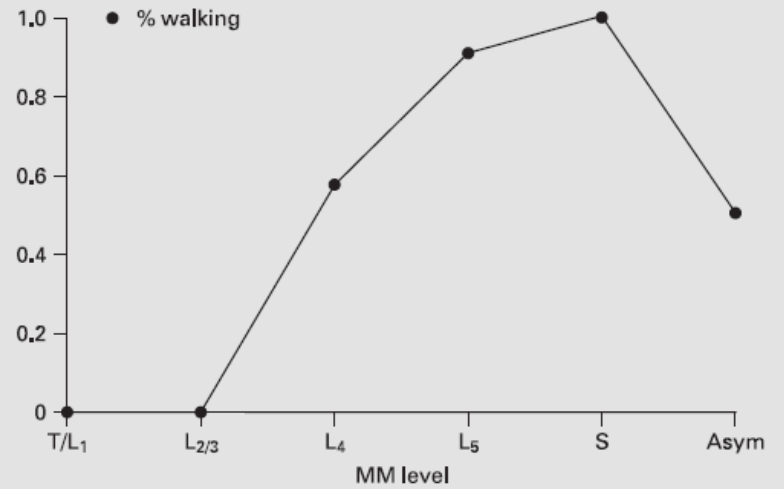
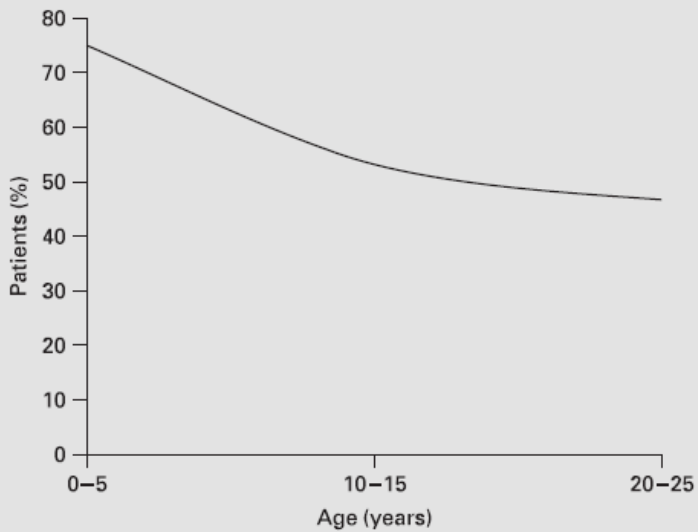


Fig. 4. This figure demonstrates the continual increase in death rate as the cohort ages.



Long term outcome

- Most are able to attend **school** although may have particular learning disabilities
- **Ambulation** possible if neurological level below L3
- Social **continence** achievable in most with CIC (Clean intermittent catheterisation)

Multi-disciplinary follow-up is the key to optimising their long term potential

- The **transition** through **adolescence** is particularly challenging as there are complex psycho-sexual issues to deal with
- They in turn will may one day have children- NB need to counsel about **folate** prophylaxis!
- Remember this condition can be prevented in most...

REVIEW

Spina bifida: A multidisciplinary perspective on a many-faceted condition

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