Clinical Guideline

SPINA BIFIDA: POST-NATAL MANAGEMENT

SETTING Southwest Region

FOR STAFF Medical or nursing staff in any hospital in the above region

PATIENTS Newborns with spina bifida

GUIDANCE

1. Background

1.1 Definitions

The neural tube usually closes between 15 and 28 days post-conception. Failure of normal closure results in a neural tube defect (NTD). This is a common congenital neurological malformation occurring in between 1-10 per 1000 births worldwide.[1] Neural tube defects can be subdivided as follows:

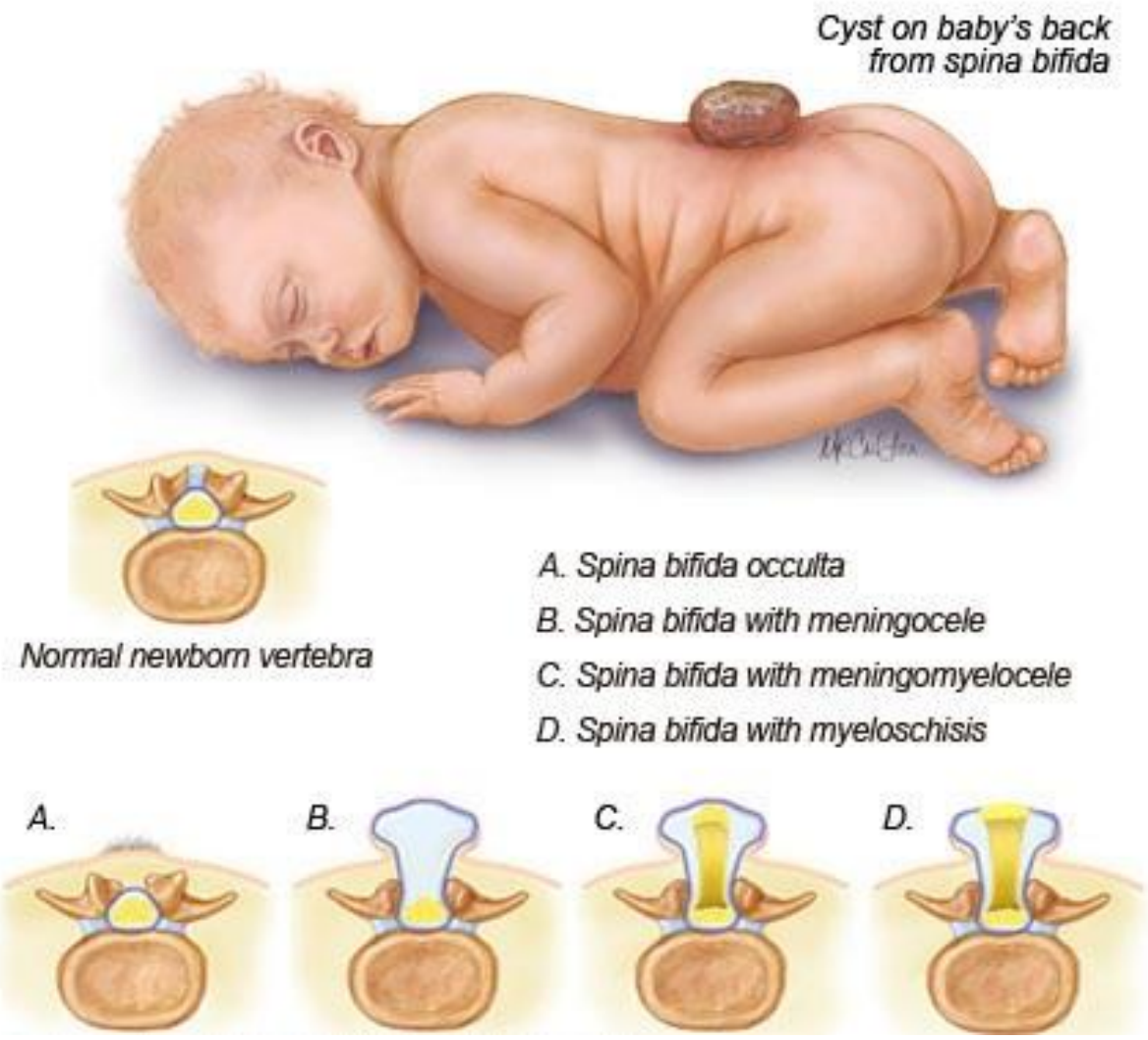
Anencephaly is caused by a defect in the closure of the anterior portion of the neural groove. This results in an unfused partially developed forebrain. It is always lethal.

Spina bifida occulta is caused by failure of closure of the posterior portion of the neural tube. It is a common anomaly consisting of a small midline defect of the vertebral bodies without protrusion of spinal cord or meninges. In the vast majority of cases this is asymptomatic and of no consequence. It relationship between spina bifida and chronic back pain is controversial. The skin at the site of the lesion may be normal, or there may be hair, dimple or birthmark. An ultrasound scan should be organised to confirm the diagnosis if such a mark is found.

Myeloschisis is the most severe form of myelomeningocele. Here, there is no overlying membrane and the nervous tissue is exposed as a flattened mass. This exposure makes the baby prone to infection.

A **myelomeningocele or meningomyelocele** is a sac which protrudes through a larger defect in the vertebral arches, containing meninges, CSF and spinal cord and/or nerve roots. This type of spina bifida often results in the most severe complications. These may include spastic diplegia, incontinence and hydrocephalus. Myelomeningoceles also have an association with the Chiari II malformation (herniation of the cerebellar vermis through the foramen Magnum, with dislocation of the fourth ventricle towards the neural canal, and downward displacement of the tentorium) other abnormalities of CNS development (e.g. polymicrogyria, heterotopias and aqueduct stenosis) may also co-exist.

Meningocele here the structure contains meninges and CSF only. In this form, the vertebrae develop normally, but the meninges are forced into the gaps between the vertebrae. As the nervous system remains undamaged, individuals with meningocele are unlikely to suffer long-term health problems, although cases of tethered cord have been reported. Causes of meningocele include tumours of the sacrococcyx presacral space and Currarino syndrome. Both meningomyelocoeles and meningocoeles may occur anywhere along the spinal cord but are most common in the lumbar and sacral area.



1.2 Genetics

Most cases of spina bifida are sporadic. A small percentage of cases have been reported to run in families; however, the condition does not have a clear pattern of inheritance. A small proportion of infants may have an associated chromosomal defect (e.g. trisomy 18, 13, or single gene disorders). If suspected a referral to the genetic service should be arranged, and karyotype/array CGH considered.

2. Post-natal management of myelomeningocele / myeloschisis

2.1 Aims of post-natal management of myelomeningocele

- Aim for surgical repair within 48 hours of delivery. Early surgery has been shown to have a lower length of hospital stay, duration of antibiotic therapy and complication rate.[2] Furthermore studies have shown early timing of surgery improves long term prognosis of neuropathic bladder.[3] Intrauterine repair is now being offered in some surgical centres worldwide, but not currently in the Bristol area.
- Latex-free handling of baby**.** This is now recognized as an important health issue for individuals with myelomeningocele. Chronic exposure to latex products from an early age in these patients has led to high rates of latex allergy with an incidence rate of at least 20% cited. Individuals allergic to latex can have life-threatening reactions to products containing latex. As such a policy of latex exclusion has been adopted for these patients. This has been shown to be effective in reducing latex allergy at a later age.[4]
- Early identification and management of urological complications of spina bifida has been shown to correlate with preservation of kidney function later in life. This includes early clean intermittent catherisation (CIC).
- Open myelomeningocele and myeloschisis should be handled sterilely with measures to ensure that the defect is kept moist and not under pressure.
- Early identification of associated conditions and complications of defect including hydrocephalus, congenital hip dysplasia and talipes equinovarus.

2.2 Delivery

1. All deliveries should be attended by neonatal team.
2. Handle baby in sterile conditions within the resuscitaire with gloves, sheets, blankets and gown.
3. Latex-free sterile gloves to be used by all team members handling the baby.
4. The baby must be placed in a side or prone position at all times.
5. The defect should be examined for size, position, and presence of neural tissue. This should be described and documented in the notes. An examination proforma and dermatome map can be found in appendix 2.
6. An open defect must be covered with sterile non-adherent gauze soaked in normal saline. A butterfly cannula (needle removed) can be placed between layers of gauze to irrigate the dressing. With its tube taped to the skin. The dressing should then be wrapped with Clingfilm. **NB it should be not applied under pressure.**
7. The dressing should not cover the anus to avoid faecal contamination.
8. All babies with a myelomeningocele should be admitted to NICU.

2.3 NICU

1. If open defect is present the baby should be nursed in a covered cot or incubator to prevent drying out of exposed defect.
2. The baby should be nursed in a prone or side position to avoid excess pressure on the defect.
3. Open defects should be dressed at described in 2.2.
4. Defect should be examined daily by a neurosurgeon.
5. Perform cranial ultrasound scan with measurement of baseline ventricular index.
6. Perform a renal ultrasound scan
7. Measure the head circumference and plot on growth chart.
8. The baby should be catheterised (latex-free) in the pre surgery period to prevent urinary retention.
9. Babies with myeloschisis or open myelomeningocele should be given 'triple' antibiotics (amoxicillin, gentamicin & metronidazole) pre-op and for 5/7 post-repair .
10. Prophylactic trimethoprim (2mg/kg od) should be started if antibiotics are not given as above.
11. Contact neurosurgery as soon as possible to arrange assessment and transfer for acute investigations i.e. MRI spine and head and possible surgical repair.
12. The baby should have a first day check and full neurological examination which should be documented in the notes.
13. Latex should be avoided as there is a high-risk of developing sensitivity, therefore document as 'latex-sensitive' in allergies section of drug chart

2.4 Preoperative care

1. Preoperative bloods should be taken including FBC, U+E, LFT, Ca, PO4, Mg, and clotting.
2. If surgery is to take place a unit of adult blood will need to be crossed matched for the baby and taken with the baby to the destination hospital's transfusion department for processing and labelling.
3. Venous access should be obtained prior to transfer and baby started on 60mls/kg/24h of appropriate fluid. Blood and other cultures are not required unless the baby is clinically unwell.
4. The baby should remain nil by mouth 4 hours prior to operation.

2.5 Postoperative Care

Urological management

1. Urology team should be contacted and informed of delivery as soon as possible. They should be given details of the patient and informed of dates for ultrasound scanning.
2. Prophylactic trimethoprim (2mg/kg od) should be started once the 'triple' antibiotics have been stopped
3. The urinary catheter should remain in situ postoperatively until the baby has been seen by the urology nurse specialist (see summary document).
4. Clean intermittent catheterisation will be established in all babies by the urology nurse specialist.
5. The baby should be discharged on prophylactic trimethoprim (2mg/kg od).
6. A follow up MCUG should be arranged for 6/52 and renal/bladder US at 2-3 months.

Hydrocephalus

1. Babies with myelomeningocele will require MRI head and spine. This is usually performed around the time of surgery.
2. The baby will require head circumference measurement and cranial ultrasound scanning (including ventricular index) completed and plotted twice weekly as a minimum.
3. Concerns of raised intracranial pressure (clinical signs such as apnoea, bradycardia, hypoventilation, and stridor and swallow dysfunction), increasing ventricular size or head circumference should be discussed with the consultant on call and neurosurgery informed.

2.6 Follow up

1. Babies should be seen by the paediatric neurosurgical nurse practitioner (NNP) at 1/52 post-op and have paediatric urology nurse specialist review in 1-2/52
2. Local/Bristol babies should have a named community paediatrician before discharge and clinic review in 4-6 weeks. Urgent referral should be made to Community Children's Health Partnership using the CCHP referral form (see Related Documents) prior to discharge.
3. Local/Bristol babies may need additional neonatal follow up in 4-6 weeks if there are specific neonatal concerns.
4. Regional babies should be referred to a named community paediatrician using their usual referral pathways with clinic review in 4-6 weeks
5. A urology appointment should be made for 3 months post discharge with ultrasound being completed prior to appointment as above. It may be possible for the baby to be seen locally (see Appendix 3).
6. Neurosurgeons should be contacted prior to discharge to discuss when they would like the baby next seen in clinic – this is usually around 6/52.
7. A referral for hip ultrasound screening for congenital hip dysplasia should be completed due to the association between congenital hip dysplasia and spina bifida. A referral to orthopaedics should be made after the ultrasound if there is evidence of hip dysplasia.

2.7 Low and high risk factors

Low Risk	High Risk
Continuous dribbling; negligible volume on CIC	*Dry interval; significant volume on CIC*
Ultrasound: normal upper tracts +/- empty bladder	*Ultrasound: pelvi-calyceal or ureteric dilatation +/- full bladder*
MCUG: no VUR	*MCUG: VUR +/-small trabeculated bladder*
No UTI's	*UTI's*
Normal bowels	*Constipation*

Appendix 1: Post-natal management of Myelomeningocele Flow Chart

Delivery
- Latex-free handling of the baby
- Sterile conditions
- Place prone or on side
- Apply moist sterile dressing to open defect

↓

NICU
- Nurse in closed cot/incubator
- Contact neurosurgical team
- Latex-free catheter
- Start 'triple' antibiotics (amoxicillin, gentamicin & metronidazole) if open defect
- Renal/bladder ultrasound
- 1st day check/neurological exam (use proforma)

↓

MRI (if indicated) & Surgery (<48 hours)

↓

Post op
- Nurse prone or on side. Daily review of wound
- Indwelling catheter to stay on free drainage until clean intermittent catheterisation (CIC) started by urology nurse specialist (on **ALL** babies)
- Prophylactic trimethoprim (2mg/kg od) once 'triple' antibiotics stopped at 5/7
- Cranial US/ head circumference measurement twice weekly. Observe for signs of raised ICP

↓

Prior to discharge
- Identify named Paediatric Urology nurse specialist
- Identify named Community or Regional Paediatrician
- Identify named Paediatric Urologist via urology registrars [bleep 5171 or 2175] – see section 2.6.4
- Ensure all follow up OP appointments arranged
- Ensure out-patient scans booked: MCUG @ 6/52, hip US @ 6-8/52 & renal/bladder US at 2-3/12

↓

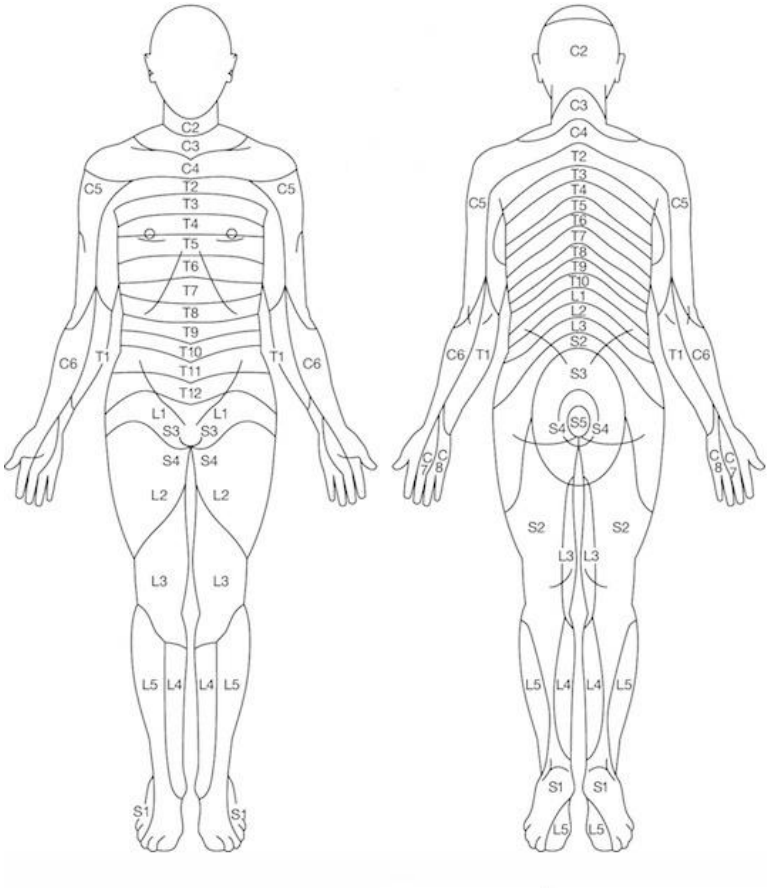
Follow up
- Paediatric Neurosurgical nurse specialist (NNP): review 1/52
- Paediatric Urology nurse specialist: review 1-2/52 re CIC
- Community Paediatrician: review 4-6/52
- Paediatric Neurosurgeon: review at approx. 6/52
- Paediatric Orthopaedic: follow up only reqd if dysplasia on routine hip scan at 6-8/52 or significant talipes
- Paediatric Urology: review at approx. 3/12

Appendix 2: Myelomeningocele Examination Proforma

Date / /
Name……..
Hospital number…….
DOB / /
Pre/post repair. Date of surgery: / /
Appearance of defect:

Neurological examination:

	Upper limb Left	Right	Lower limb Left	Right
Tone				
Power				
Reflexes				
Sensation (see dermatome chart)				



Appendix 3: References

1. Epidemiologic and genetic aspects of spina bifida and other neural tube defects. Au KS, Ashley-Koch A, Northrup H. Dev Disabil Res Rev. 2010;16(1):6-15.
2. The effect of surgery time on prognosis in newborns with meningomyelocele. Oncel MY, Ozdemir R, Kahilogulları G, Yurttutan S, Erdeve O, Dilmen U. J Korean Neurosurg Soc. 2012 Jun;51(6):359-62.
3. The timing of primary neurosurgical repair significantly affects neurogenic bladder prognosis in children with myelomeningocele. Tarcan T, et al. J Urol. 2006 Sep;176(3):1161-5.
4. Children at risk: latex allergy and spina bifida. Leger RR, Meeropol E. J Pediatr Nurs. 1992 Dec;7(6):371-6.

Appendix 3: Discharge check list

	NAME	Review: when and where?
NEUROSURGICAL NURSE PRACTITIONER (NNP):		
PAEDIATRIC UROLOGY NURSE SPECIALIST:		
COMMUNITY PAEDIATRICIAN:		
PAEDIATRIC NEUROSURGEON:		
PAEDIATRIC UROLOGIST:		

	DATE	Where?
MCUG		
HIP ULTRASOUND		
RENAL TRACT ULTRASOUND		

Paediatric urology follow up

This will normally be arranged by the urology registrars according to area covered below.

Consultant Paediatric Urologists [*area covered*]:

i. [redacted] [**LEAD**; *Bristol, Swindon, Yeovil*]
ii. [redacted] [*Barnstaple, Gloucester, Torbay, Truro, Plymouth*]
iii. [redacted] [*Exeter, Gloucester, Taunton*]

If follow up arrangements aren't clear, then contact [redacted]

Draft guidline originally created by Richard Lee-Kelland, Paediatric trainee

RELATED DOCUMENTS CCHP Single Point of Entry Referral Form [redacted]

SAFETY No issues

QUERIES Contact [redacted] or his secretary on [redacted]