Nursing care and surgical correction of neonatal myelomeningocele

Neural tube defects are a major cause of fetal, neonatal and childhood death. They are also responsible for significant life-long disabilities. In some cases, they can be prevented, and in others surgery can improve life expectancy and quality. Factors relating to these concepts are discussed, alongside suggestions for improvement in care.

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Keywords

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Key points

1. Periconceptional folic acid supplementation is known to reduce neural tube defect incidence but the impact on incidence within the general population appears negligible.
2. Surgical correction of spina bifida – myelomeningocele – is improving, reducing mortality and morbidity.
3. Neonatal surgical care utilises basic but important actions, improving immediate and long-term outcomes.
4. National guidelines on neonatal neurological assessment are lacking.

Congenital abnormalities are described as anomalies involving structure, metabolic or endocrine processes, or genetic coding that is present in the newborn at the end of the pregnancy[1]. Neural tube defects are the most common abnormality of the central nervous system, probably second only to cardiac defects when considering major congenital anomalies[2]. Representing a major public health concern, neural tube defects have a dramatic impact on all social levels by virtue of their mortality, morbidity, social cost and human suffering[2].

Embryology

Neural tube defects arise from abnormalities in the formation of the embryonic neural tube, the forerunner of the central nervous system and most of the peripheral nervous system. By day 28 of embryonic life, before most mothers are aware of their pregnancy, the neural tube has formed from the neural crest, through a complex and highly regulated morphogenetic process[3]. Approximately 132 genes have been implicated in the control of this process, with neural tube defects arising from a rostral or caudal failure of neural tube closure[4]. While the precise process of neural tube closure is not known, two models have been proposed. The first suggests a zip-like closure of the neural tube; the second proposes multiple closure sites along the developing neural tube[5]. Some defects may arise from secondary reopening of the tube or from a post-neurulation defect[6].

Types of neural tube defects

The clinical spectrum of neural tube defects spans five forms, namely, craniorachischisis, iniencephaly, anencephaly, encephalocele and spina bifida. Craniorachischisis is rare, with an incidence ranging from 0.1-10.7 in 10,000. It results in fetal or early neonatal death, and the huge variation in incidence is strongly linked to increasing levels of poverty[7]. Iniencephaly is also rare, with an incidence of 0.1-10 in 10,000, and similar outcomes[8]. The USA records the incidence of anencephaly, encephalocele and spina bifida as 1.4, 5.5, and 3.7 in 10,000 live births, respectively[5]. By comparison, and for reasons unknown, Wales has the highest recorded incidence of these three neural tube defects in Europe, with rates as high as 17 in 10,000[9]. Anencephaly will usually result in fetal or early neonatal death[3]. Encephalocele mortality is largely determined by the site and size of the lesion while early childhood survival of spina bifida can be near 100%[6].

Despite perceived negative outcomes, progress in medical treatment has improved the outcome for children with the survivable neural tube defects of encephalocele and spina bifida. Surgical correction of encephalocele can be straightforward, with a favourable prognosis, dependent on the site and extent of the lesion[6]. Substantial improvements in the surgical correction of spina bifida are also described, with mortality falling from 37% in the first year of life in 1972, to a mean survival of 30 years[6]. Of the three main forms of spina bifida (see **FIGURE 1**), literally 'split spine', only spina bifida occulta does not require surgery to correct the defect.

Spina bifida occulta is described as arising from flawed closure of the vertebral laminae, though the spinal cord and neural tissue are usually normal[10]. Although it is suggested that this is present in approximately 5% of the USA population, the symptoms experienced by the affected

individuals are usually so mild they are unaware of the defect. In contrast the two forms of spine bifida cystica – meningocele and myelomeningocele – both require surgical intervention[10]. In both forms, the meninges protrude through the malformed spinal column. In myelomeningocele, the most common form and that which is usually referred to as spina bifida, this is further complicated by the presence of neural tissue. After corrective surgery for myelomeningocele (see **FIGURE 2**), there is always neural insufficiency to regions below the defect, which may manifest in conditions such as paraparesis and neurogenic bladder and bowel[10]. However, there is usually little or no neural damage and disability after surgery to correct meningocele.

While such physical disabilities and their life-long implications are important to remember, anomalous brain development causes its own concerns. Hydrocephalus, with its associated functional and neural abnormalities, occurs alongside myelomeningocele in up to 90% of cases, with up to 80% historically requiring surgical intervention for this condition[11]. Current trends indicate that this figure is now nearing 50%[12]. Differing degrees of Chiari II malformations, with significant impact on the hindbrain and brainstem, occur in virtually all cases of myelomeningocele and can give rise to diverse symptoms including apnoea, headaches and bradycardia[13].

Folic acid and the prevention of neural tube defects

Precise causative agents for neural tube defects are largely unknown[14], though many factors have been implicated. These have included socioeconomic status, maternal age, birth order, parental occupation, maternal medication and caffeine use, and even hypothermia in early pregnancy[4]. Furthermore, there is a 50-fold increased risk for recurrence of neural tube defects in subsequent pregnancies[15]. Nonetheless, the complex aetiology of neural tube defects appears to be substantially influenced by maternal diet[3]. Seminal work by Smithells et al[16] and Laurence et al[17] provided two of the first studies demonstrating conclusive links between poor maternal diet, specifically a deficiency of vitamin B9 (folic acid), and an increased neural tube defect incidence. Studies such as these, culminating in the randomised double-blind prevention trial of the Medical Research Council Vitamin Study Group[18], prompted government advice on the periconceptional use of folic acid[19]. Women of childbearing age were advised to take 400µg/day before and during early pregnancy; those with a family history of neural tube defects, 4mg/day.

Despite the common perception of folic acid being the panacea of neural tube defects and contemporary literature citing its undoubted benefits[3], others state that periconceptional use does not appear to have fulfilled its perceived potential. Decreasing neural tube defect incidence in England and Wales has been noted since the early 1970s, and folic acid supplementation has had no discernible impact on this downward trend[20]. A similar situation is described in both the USA and many, mainly western, European countries[2] although it may be possible to attribute these findings to nearly 50% of pregnancies being unplanned and lack of knowledge about the use of folic acid supplementation[2,20]. These findings led to universal fortification of the diet with folic acid in the USA, with some fall in neural tube defect incidence[2]. However, this measure was rejected in the UK over fears that it would mask vitamin B12 deficiency in the elderly[9]. Inborn maternal[3] and fetal[21] errors of folate metabolism have also acted to reduce the efficacy of folic acid supplementation.

Antenatal screening

Concurrently, antenatal screening in the UK has improved dramatically, moving from reliance on low-quality ultrasonography and blood serum tests to indicate the possibility of neural tube defects with a wide regional variation in screening protocols[22], to high-quality cost-effective ultrasonography wielded by highly experienced health professionals[23]. A review of ultrasonography use in the UK reports a near 100% detection rate of anencephaly by week 14, and 66% detection rate for spina bifida in the same time scale[23]. Similar success rates have been reported in Wales[9]. In the USA, such improvement in antenatal screening over the past 30 years has also increased the prevalence of selective termination of affected pregnancies, further increasing the difficulty in determining the effectiveness of folate supplementation, as incidence is usually determined by the number of live births[2]. By comparison, studies found that between 1998-2009, while the incidence of spina bifida in Wales fell, determined by antenatal diagnosis, the number of affected live births rose, indicating an active choice to continue with affected pregnancies[24].

Impact on families

While antenatal screening for neural tube defects has significantly improved, there is a lack of input from those directly affected by such advances – the parents[25]. An Australian study explored the feelings and emotions experienced after antenatal diagnosis of a neural tube defect or hydrocephalus[25]. Portraying recognised patterns of grief and bereavement, these parents at once feel helplessness, disorientation and shock[26]. It was found that, despite determination to continue with the pregnancy, such feelings could persist for the duration of the pregnancy[25]. While admission to a neonatal unit is expected, the experience is stressful[27], and such feelings are only heightened in the milieu of the neonatal unit[28]. Consistent, empathetic support by neonatal staff can begin to provide appropriate support for these parents[29]. Alongside these initial stressors, parents must now also consider the implications of providing life-long care for a child who may have severe physical
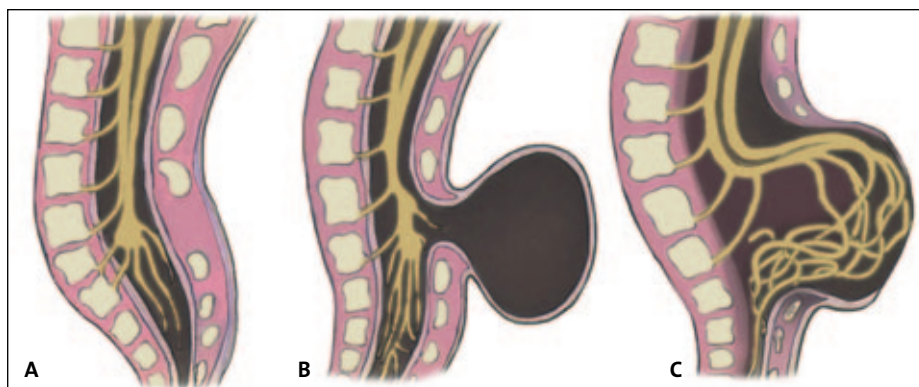
FIGURE 1 Three types of spina bifida. A) Spina bifida occulta; B) Meningocele; C) Myelomeningocele.
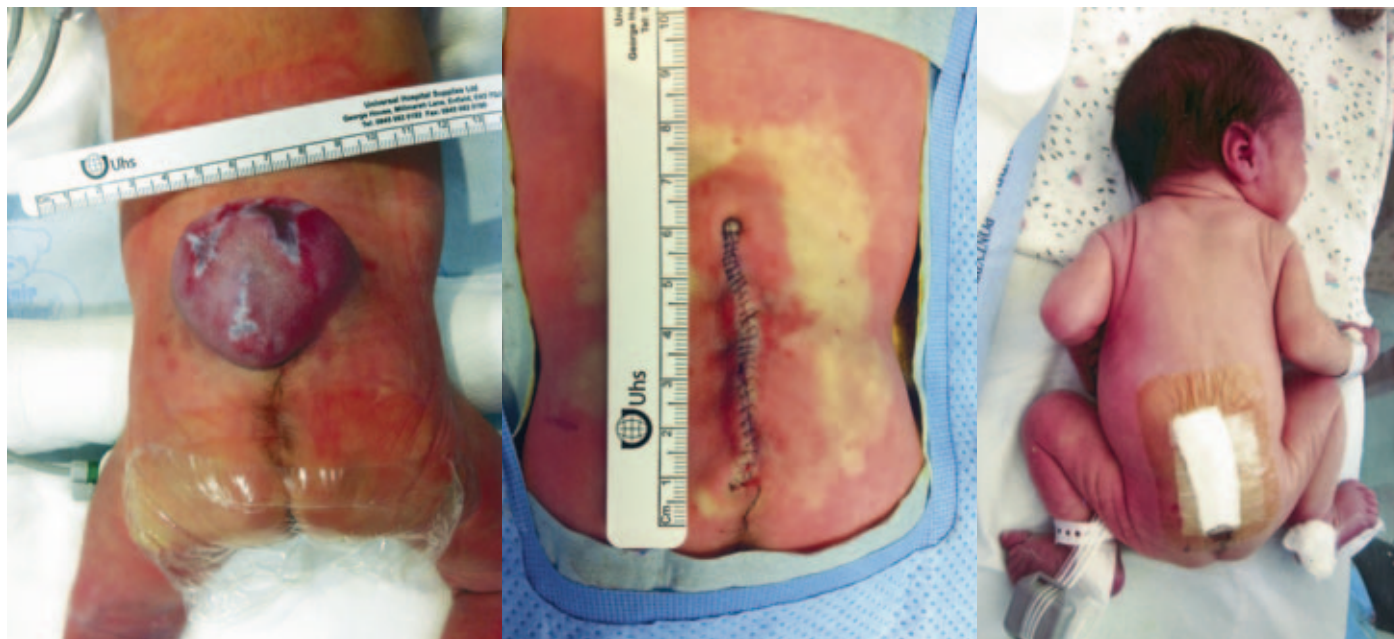
FIGURE 2 Corrective surgery for myelomeningocele.

and mental disabilities. Stressors on such families can lead to global psychological distress, including stress and depression, in both parents. Siblings suffer in a similar manner, also reporting great concern for the isolation of the affected child, and sadness that their sibling is not able to engage in physical activities. In contrast, affected families can also demonstrate great resilience and develop much stronger family bonds through this experience[30]. Formation of good therapeutic relationships with neonatal staff can pave the way for future health professionals, enabling them to improve the support of the family[31].

Perinatal management of neural tube defects

While the optimal mode of birth in antenatally diagnosed myelomeningocele is debatable[6,32], it was thought that myelomeningocele repair should occur soon after birth[33]. Such prompt intervention was thought to help reduce infection risks and preserve neural function. Once considered a surgical emergency requiring immediate attention, there is little evidence to support this viewpoint[6]. Stabilisation of the infant is more important and clinically this entails achieving stable and appropriate vital signs, ensuring adequate respiration, pulse rate and temperature, and usually including mean blood pressure[34]. While the infant is stabilised, it is vital that the lesion is examined, noting location and possible leakage of cerebrospinal fluid[32]. A thorough

neurological examination is also useful, noting aspects such as spontaneous activity, extent of muscle weakness/paralysis, orthopaedic deformities – limbs or spinal, and the presence of hydrocephalus or Chiari II malformation[32]. A concurrent assessment for associated congenital anomalies, including cardiac and renal irregularities[32] will also improve subsequent care. Early proactive management of these anomalies can significantly improve future quality of life[35,36].

Nursing care and surgical correction of myelomeningocele

The main surgical goals in the repair of myeolomeningocele are the formation of the neural tube and skin closure[32]. There may also be attempts to repair the vertebral defect. In the preoperative period, there are four basic principles of nursing care to consider for affected neonates[33]. The first of these is the use of sterile techniques when caring for the defect, which is an open track for pathogens to the central nervous system. Second, is the prevention of hypothermia. While all neonates are at risk of hypothermia[37], the open defect may make thermal homeostasis more difficult to maintain. This difficulty arises because the skin covering of most myelomeningoceles is incomplete[38]. As such, unprotected internal structures are exposed to the environment, increasing radiant heat loss. Also, leaking cerebrospinal fluid will increase evaporative heat loss. Both of these factors will tend to cool the neonate, and may

necessitate active management to maintain appropriate body temperature. The use of plastic wrap may reduce the impact of these routes on thermoregulation[32]. The third principle is to nurse the neonate prone, with the defect covered in saline moistened dressings. The exposed neural tissue is potentially functional, and so must be preserved[31]. These actions seek to preserve potential neural function by avoiding mechanical damage and desiccation of the exposed neural tissue[33]; plastic wrap can also improve the efficacy of the saline dressings[31].

Finally, avoiding the use of latex as these neonates are at increased risk of developing latex sensitivity[33]. Latex allergy incidence in these children can be as high as 25-65%, compared to 0.7% in unaffected children. It is direct latex contact with the meninges and mucosal membranes in early, frequent surgeries and procedures which is key to this prevalence[39]. Altered neuroimmune interactions may also predispose this population to latex sensitivity[38]. Hence, latex-free environments reduce sensitisation episodes and the subsequent risks of anaphylactic reactions. The use of broad-spectrum prophylactic antibiotics appears to be supported by a reduction in postoperative infection[32].

Building on antenatal counselling, parents should be prepared for imminent surgery. The possibility of immediate, intermediate and long-term complications should be discussed. These may include hydrocephalus[11], complications related to cerebrospinal fluid shunt placement[40],

neurogenic bladder[35], developmental delay[41] and orthopaedic difficulties that may include paraplegia or paraparesis[36].

A pioneering surgical option being explored to correct myelomeningocele is prenatal surgery[32,42]. Performed at around 26 weeks' gestational age, reports currently suggest some long-term benefits for the child. These include reduced need for cerebral spinal fluid diversion, reduced incidence of Chiari II malformation and improved motor outcome. However, the oldest patients are currently only 30 months old. Clinical experience suggests that similar results are seen in patients in receipt of postnatal repair, whose neurological function then deteriorates with age. It has yet to be seen if this occurs in prenatal surgical patients. Furthermore, it is highlighted that the procedure also carries significant risks for both the mother and fetus, including extreme preterm delivery, uterine dehiscence at delivery, and placental abruption. While this work is promising, it is acknowledged that further study is required to develop this into a viable treatment option.

Postsurgical care

Following postnatal myelomeningocele repair, many infants will develop some degree of ventriculomegaly[32]. This arises from imperfect cerebrospinal fluid circulation attempting to follow conduits that have not been established *in utero*[38], causing head circumference to increase at a rate greater than normal[32]. Assessment of ventriculomegaly and associated hydrocephalus can be made by serial ultrasound scans[32]. Combined clinical and radiographic assessment, including head circumference measurements, can be used to determine the need for cerebrospinal fluid diversion by means of a shunt[32]. Prompt action avoids issues of mortality and morbidity associated with long-term hydrocephalus[11]. However, infants with stable neurological status and stable or slowly increasing ventricular size, need only clinical assessment, with regular head circumference measurements and imaging studies[32]. This practice reduces the frequency of shunt placement and avoids its associated long-term complications[40].

Benchmarking with a number of UK level 3 neonatal units highlights a range of methods utilised for clinical neurological evaluation after myelomeningocele repair. These methods vary from the full Glasgow Coma Scale[43] to simple observations recorded in narrative form that may or may not include pupil reaction. In clinical experience, while the observations made can be useful, the impetus for recording postoperative neurological observations appears to arise more from routine practice rather than the needs of the neonate. A similar situation is described in an Australian study into drivers for postoperative observations[34]. While acknowledging the need for postoperative observations, it is emphasised that these need to be driven by the needs of the patient, using an evidence-based format[34].

It is necessary, therefore, to have an evidence-based assessment tool with which to make such clinical judgements. The use of a standardised assessment tool not only improves reliability of nursing records, but also increases the accuracy of notes that are traditionally written in narrative form[44]. Accurate and standardised evaluation is vital in the assessment of neonatal neurologic status, which must take into account not only the maturity of this rapidly developing system but also potential pathological changes[45]. However, national guidance on this issue is lacking. The National Institute for Health and Clinical Excellence (NICE)[46] only has guidelines for neurological assessment of infants and children who have suffered head trauma, but none for these age groups relating to neurological evaluation following neurosurgery. Furthermore, while tools for evaluating neurologic status in newborns exist, they tend to be detailed examinations requiring the baby to be held and moved, and so are more suited to discharge checks and prognosis of future neurological condition[45,47]. Additionally, while recognising neurological evaluation is important, the needs of the neonate as a whole must not be forgotten. Care delivery models can improve holistic nursing, including wound care[48]. This supports the observation that standardised tools improve the reliability and accuracy of nursing notes[44].

It is recognised that the life-long implications of neural tube defects are significant, both for surviving children and families. It appears that significant improvement can be made in public health arenas regarding the periconceptional use of folic acid. Increased public compliance with folic acid supplementation may reduce the incidence of neural tube defects still further. However, it is conceded that not all neural tube defects can be prevented in this manner. Hence, in partnership with improvements in postnatal surgical repair of myelomeningocele, it is suggested that developments are also encouraged in postoperative nursing care. The mainstay of this would be a standardised evidence-based tool that would support effective holistic evaluation of the neonate. With such care, it is hoped that the quality of life for affected children will be improved.

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