

# Neural tube defects in newborns

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## ABSTRACT

**Objectives:** The aim of this study was to review cases of neural tube defect with special focus on presentation, epidemiology and clinical presentation. Management with complications and results of surgery were discussed.

**Methods:** Ninety-three cases, born with neural tube defect (spina bifida aperta) were operated upon in our unit at King Hussein Medical Centre, Amman, Jordan, from June 1997 to October 2000. Data was retrospectively reviewed and analyzed.

**Results:** Major neurological deficits were present in 28 cases (complete loss of function below the level of the lesion) and 19 cases were intact. Lesion size was less than 3.5 cm in 72 cases (77.4%); 3.5-7.5 cm in 15 cases (16.1%) and 6 cases were more than 7.5 cm. Seventy-two cases had an intact lesion before surgery while the other 21 cases ruptured either during delivery or soon later. For the site of lesion, 51 cases were lumbar, 22 cases were lumbosacral, 13 cases were thoracolumbar, 4 cases were thoracic and 3 cases were cervical. Early surgery was carried out for all cases, primary closure was possible in all cases. Nineteen cases had post-operative complications, wound infection (superficial) 6 cases (7%), leakage of cerebrospinal fluid 5 cases (5%), meningitis 3 cases (3%), skin necrosis 3 cases (3%) and 2 deaths.

**Conclusions:** Jordan has a large number of born spina bifida cases, as the practice of pregnancy termination is socially and religiously unacceptable. Our approach to management, is to repair all intact patients. For patients with major neurological deficits we advise the family on the natural history of the disease and postoperative status of the patient leaving the decision to the family. The presentation was similar to that described in literature.

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Neural tube defects are a common problem encountered at King Hussein Medical Centre, Amman, Jordan, and despite advanced and appropriate prenatal care we still deal with a considerable number of cases. This is related to the fact that termination is not an acceptable practice due to social and religious basis. Neural tube defects are classified into spina bifida occulta and aperta. Both are characterized by congenital absence of spinous process and variable amount of lamina, in spina bifida occulta there is no visible exposure of meninges or neural tissue.<sup>1</sup> Spina bifida aperta is divided into 2 entities according to the tissue exposed. They are meningocele, where there is cystic distension of the meninges with no abnormality of neural tissue and myelomeningocele, where the cystic distension of the meninges is associated with functional and structural abnormality of spinal cord or cauda equina.<sup>2</sup> In this study we have focused on cases with spina bifida aperta (meningocele and meningomyelocele).

Methods. Ninety-three cases, 58 females and 35 males, born with neural tube defects were operated upon in our unit at King Hussein Medical Centre, Amman, Jordan from June 1997 to October 2000. Data such as prenatal history, radiological findings, associated anomalies, neurological findings, size and site of

lesion and presence of hydrocephalus were reviewed and analyzed. All patients were received within the first 72 hours after birth, 83 patients (90%) within the first 24 hours after birth and 88 patients (95%) were diagnosed antenatally. Ultrasound (US) was the primary diagnostic tool as it was used in all cases, only 6 patients had serum alpha fetoprotein (AFP) values assessed; it was raised in all 6 cases. None had amniotic AFP analysis. Vaginal delivery was the case in all patients except for 10 patients where cesarean section was performed for obstetric indications, Table 1. All patients were operated upon as soon as they had been received and prepared for surgery, mean time was 24 hours, ranging from a few hours to one week. Ruptured cases were started on antibiotics as soon as they were received or the rupture occurred, no antibiotics were used for the non-ruptured cases. Ceftriaxone (Rocephin) was the drug used. It was stopped 3 days after the repair was achieved. Brain US was used for the diagnosis of hydrocephalus, computerized tomography (CT) scan of brain was carried out preoperatively for all patients undergoing shunting.

**Results.** Hydrocephalus was found in 30 (32%) cases at presentation and they were shunted at the same time of repair, another 40 (43%) patients developed hydrocephalus within 2-4 weeks from the time of repair, mean time of 10 days. In Tables 2-7 we summarize the patients' presentation, lesion size, spinal level, nature of lesion cover, radiological findings and associated congenital anomalies. Surgery was performed under general endotracheal anesthesia in prone position, antibiotics were given in all cases at induction and 3 doses after. Primary closure was achieved in all cases, release incisions were needed in 5 cases. Plastic surgeons assistance were needed in 3 cases where flaps were used, these cases were of the big defect group (>7.5 cm). The filum terminale was identified and resected in 32 cases, the other cases we could not identify the filum or it was more damaging to try to locate it. The neural placode in meningomyeloceles were dissected free from the surrounding tissues and imbedded within the repaired, refashioned dural tube. Cases diagnosed to have hydrocephalus at the time of surgery were shunted in the same set up. Postoperative complications occurred in 19 cases (20%), wound infection in 6 cases (7%) leakage of cerebrospinal fluid in 5 cases (5%), meningitis in 3 cases (3%), all cases were due to gram negative microorganisms. Skin necrosis 3 cases (3%) and death 2 cases (2%), both were due to wound infection and meningitis (3 and 7 days after diagnosis of meningitis) Follow up post operatively showed deterioration in pre-operative neurological findings in 2 cases, the rest remained unchanged.

**Discussion.** Neural tube defects resulting from failure of the neural tube to close during the 4th week of embryogenesis are the most common severely disabling birth defects in Jordan. Depending on the level of the lesion, interruption of the spinal cord at the site of the spina bifida defect causes paralysis of the legs, incontinence of urine and feces, anesthesia of the skin and abnormalities of the hips, knees, and feet.<sup>3</sup> We deal with a large number of born spina bifida cases, as the practice of pregnancy termination is socially and religiously unacceptable. Despite adequate and proper antenatal care and periconceptional folic acid use, we still deal with what we feel is an unacceptable high number of cases. No definitive statistics are available as to the incidence of spina bifida in Jordan, but as almost all patients diagnosed intrauterine reach term and are delivered, we still face a large number of cases. United States of America values are one in every 2000.<sup>4-6</sup> We repair all intact patients unless medically contra indicated. For patients with neurological deficits we advise the family on the natural history of the disease and postoperative status of the patient leaving the decision to the family.<sup>7-10</sup> The presenting signs and symptoms were nearly identical to that described in literature. Repair of the defect did not affect the neurological findings in terms of improvement.<sup>11,12</sup> Two cases had deeper weakness after surgery, both resulting from pre-operative injury to nervous tissue (placode in one and

root in the other).

Finally, spina bifida is a challenging problem in Jordan, despite advanced health care services and maternity care. We need to work on reducing numbers of new cases, which does not seem to be an achievable goal in the near future. Generally, we discourage close relative marriages especially in cases with positive family history. Folic acid is now a routine practice for obstetricians. Genetic counselling is available when needed.

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