Background: The long-term prognosis of patients with Spinal Open Neural Tube Defect (SONTD)-associated hydrocephalus is not well known. This study was conducted to ascertain the incidence and natural history of hydrocephalus in patients with SONTD.

Methods: All 82 patients with SONTD referred to Neurosurgery/Spina Bifida Clinics at King Khalid University Hospital, Riyadh, Saudi Arabia (January 1995 - July 2010) were studied and followed for a period of 1-16 years. Patients were divided into three groups: Group “A” with active hydrocephalus treated with ventriculoperitoneal shunt (VPS), or endoscopic third ventriculostomy (ETV); Group “B” with compensated hydrocephalus; and Group “C” with no hydrocephalus. Timing of shunt insertion, complications of treatment and status of hydrocephalus were analyzed.

Results: The mean age of the 82 patients was 7.4 years (range 1-16 years). Group “A” included 59 (72%) patients, Group “B” 7 (8.5%) patients, and Group “C” 16 (19.5%) patients. Chiari malformation type II was found in 71 (86.6%) patients, 57 of whom (80%) were in Group “A” with active hydrocephalus. They were treated by VPS (51 patients) and ETV (8 patients). The shunts were revised or replaced in 10 (19.6%) patients due to obstruction or infection. Primary ETV failed in 3/8 patients, and treated by VPS. None of those in Groups “B” or “C” required treatment for hydrocephalus during the follow up.

Conclusion: Hydrocephalus affects the majority of patients with SONTD who have Myelomeningocele (MMC) and CM II and requires close surveillance and prompt management. Children with SONTD should routinely undergo MRI examination of brain and craniocervical junction to clarify ventricular size, and the presence of CM II.

Key Words: Chiari malformation type II, hydrocephalus, myelomeningocele, spinal open neural tube defect

INTRODUCTION

Although the exact incidence of hydrocephalus in neural tube defect (NTD) is not known, it has often been considered an almost inevitable sequela of spinal open neural tube defect (SONTD). It may be absent at birth but develops in the first weeks or months of life.[22] A variety of factors are implicated in the pathogenesis of
hydrocephalus in these children; aqueductal stenosis and Chiari malformation type II (CM II) which is characterized by herniation of the cerebellar vermis, and fourth ventricle through the foramen magnum, with kinking of the brainstem, beaking of the tectum, and a small posterior cranial fossa” [Figure 1].[8] Hydrocephalus usually develops secondary to impaction of the posterior fossa contents on the foramen magnum, leading to occlusion of the outlets of the fourth ventricle with cerebrospinal fluid (CSF) outflow blocked, or impaired, at the foramina of Luschka and Magendie and resulting in progressive ventriculomegaly. Moreover, vertical translocation of the brain stem causes increased resistance of CSF flow through the tentorial hiatus. The small volume of the posterior fossa, in conjunction with the very abnormal tilt of the tentorium, and cerebellar prolapse causes increased tension inside the posterior fossa, which leads to increased resistance of the venous outflow through the sigmoid sinus and venous hypertension. This may create an element of communicating hydrocephalus.[18,19]

There is a high association between SONTD and CM II.[18,19] This association is unique to MMC and is found only in this population of NTD patients. Hans Chiari described, in 1891 seven patients with combination of hydrocephalus and spina bifida, at ages ranging from birth to 6 months. However, priority in description of the CM II does not go to Chiari. Rather, this description was first published by John Cleland in 1883, some eight years before Chiari.[2,3]

Multiple ventricular anomalies are commonly found in CM II patients. The fourth ventricle, is typically small and poorly visualized, and is frequently displaced into the cervical canal along with its choroid plexus. The aqueduct is similarly small and rarely seen on routine imaging, but this might not contribute significantly to hydrocephalus [Figure 2].[17] Third ventricle is rarely enlarged but may take a narrow-angled appearance, giving rise to the term “shark tooth deformity”. The lateral ventricle varies from nearly normal to being severely deformed and hydrocephalic. Colpocephaly is common with the occipital horns disproportionately enlarged compared with the frontal horns [Figure 3]. This finding is often present even in patients with MMC who do not have hydrocephalus and frequently persists in patients in whom a shunt has been placed [Figure 4].[8,16]

The predominant treatment of hydrocephalus continues to be CSF diversion, most commonly into the peritoneal cavity by means of ventriculoperitoneal shunt (VPS). However, these shunts exhibit a variety of modes of failure: the most common of which are mechanical obstruction, excessive CSF drainage, and infection. Failure rate may reach up to 40% in the first year after initial insertion.[9]

Before shunting became established, these patients had a poor prognosis, the majority were not offered treatment, and only 20% of non-operated children with hydrocephalus and MMC reaches adulthood, with poor intellectual outcomes. After the introduction of shunting, outcomes improved significantly.[4,11,21]

The life expectancy of children with MMC has improved dramatically since the introduction of modern intensive interventions in the last 45 years. Most of the deaths occur in the first year of life, mostly due to renal and respiratory problems associated with spina bifida. Only a few of the deaths are related to hydrocephalus.[4]
MATERIALS AND METHODS

The study included children with SONTD, (where spinal cord was exposed at birth through a defect in the posterior component of the spine) who were evaluated at the Spina Bifida Clinic, at King Khalid University Hospital (KKUH), Riyadh, Saudi Arabia, between January 1995 and July 2010. These patients were recruited from those who had SONTD; spina bifida, MMC and CM II malformation, with or without hydrocephalus.

All case records of children who were born in KKUH, or referred with the clinical diagnosis of SONTD, and confirmed by neuroimaging studies; computerized tomography (CT) and/or magnetic resonance imaging (MRI) were reviewed. Relevant information about each child was retrieved. This included age, sex, neurological assessment, (CT and/or MRI), timing of treatment of hydrocephalus (whether by a ventricular shunt or ETV), course of hydrocephalus, and sequelae and outcome were recorded. Exclusion criteria were cranial neural tube defect, spina bifida occulta, closed meningocele or meningo(myelo)cele, lipomeningocoele or lipomeningomyelocele, and patients lost for follow up.

Patients were divided into three groups according to the status of hydrocephalus and size of ventricular system. Group “A” were patients with active hydrocephalus, because of progressive ventricular dilation and presence of signs of high intracranial pressure (ICP). These were treated by VPS, or ETV. Group “B” included patients with compensated (arrested) hydrocephalus, with no signs of raised intracranial pressure (ICP) [Figure 5]. Group “C”: patients with no evidence of hydrocephalus [Table 1].

All patients were followed up regularly at the Spina Bifida Clinic every 6 months for assessment of shunt function and status of hydrocephalus. Causes of shunt failure, and number of revisions at the time of follow up were also analyzed.

RESULTS

A 15 - year retrospective database search of patients registered in the Spina Bifida Clinic between 1995 and 2010 yielded a total of 170 patients. Of these 82 patients with confirmed diagnosis of SONTD were evaluated. They consisted of 44 males and 38 females. The mean age at the time of the last follow up was 7.3 years (range: 1-16 years), and the mean duration of follow-up was 68 months (range: 12 to 119 months).

Chiari Malformation type II was the main cause of hydrocephalus in 71 (86.6%) patients. Fifty seven of them (80.3%) received treatment for active hydrocephalus (in two patients of Group “A”, hydrocephalus was secondary to aqueductal stenosis, and they had no evidence of CM II). All the 7 patients in Group “B” with compensated hydrocephalus had CM II, whereas 7 out of the 16 patients with no hydrocephalus in Group “C” had CM II [Table 1].

Forty two (51.2%) patients showed marked colpocephaly with enlargement of the occipital horn of the ventricles due to underdevelopment of the white matter in the...
posterior cerebrum. The appearance was of apparent hydrocephalus of occipital horns, while the frontal portions were relatively normal [Figure 3].

Group “A” consisted of 59 (72%) patients who had active hydrocephalus. All of them underwent treatment; 51 patients had VPS, and 8 patients ETV. Ventriculoperitoneal shunts were inserted simultaneously at the same time of MMC repair in 30 patients. In 16 patients, hydrocephalus was treated within one week from the MMC repair, while delayed treatment (after one week) was performed in 13 patients. No patient in this series required treatment of hydrocephalus later than 6 months of age. The 7 (8.5%) Group “B” patients were diagnosed to have compensated type of hydrocephalus and did not require treatment during follow up. The remaining 16 (19.5%) patients, who comprised Group “C”, had no hydrocephalus and remained asymptomatic [Figure 6].

Among 51 patients with VPS, 5 (9.8%) experienced at least one surgical shunt complication during the follow up period, and another 5 (9.8%) had multiple (two or more) shunt complications. Shunt replacement or removal occurred in 90% of patients with a shunt complication, whereas ETV and removal of the obstructed shunt was performed in one patient. All cases of shunt infection were seen in the first year after insertion while only two of the obstructed shunts occurred after one year of insertion. Three patients with shunted hydrocephalus developed intra ventricular cysts which became symptomatic in one patient after increasing in size and causing significant shunt blockage. He was successfully treated by endoscopic fenestration of the cyst and readjustment of the ventricular catheter [Figure 7]. Primary ETV failed in 3/8 patients within 6 months of the ETV. The reason of failure was not known as those patients were treated by insertion of VP shunt on emergency basis. Two patients died due to complications related to hydrocephalus; shunt failure, repeated revisions and respiratory failure.

DISCUSSION

The term SONTD is used to describe a defect that occurs when there is failure of fusion of the neural tube and herniation of meninges with or without neural elements, lacking skin covering or covered by a thin transparent membrane that does not prevent CSF leak and exposing neural structures. The incidence of MMC ranges between 0.2 and 2.0 per 1000 live births, with regional and racial variations. The overall incidence has declined significantly in the last 2 decades, due to improved maternal nutrition during pregnancy with addition of folic acid, and therapeutic termination of affected pregnancies. Exact incidence of hydrocephalus in SONTD is not known. In a significant proportion of patients, hydrocephalus is absent at birth but develops in the first few weeks or months of life.

Ventriculomegaly was seen in patients with MMC even before closure of the defect in 15-25%, but in most surgical series the proportion of children with SONTD-associated hydrocephalus who require shunting reaches up to 80%-90%. Steinbok et al. found 84% (85/101) of children with MMC required VPS, and Rintoul et al. reported 81% of affected individuals required ventricular shunting. In the present series 80.5% (66/82) of patients

| Group* | A | B | C | Total (%) |
|--------|---|---|---|-----------|
| No     | 59 | 7 | 16 | 82 (100)  |
| Male   | 31 | 5 | 8  | 44 (53.7) |
| Female | 28 | 2 | 8  | 38 (46.3) |
| CM II  | 57 | 7 | 7  | 71 (86.6) |
| Hydrocephalus | 59 | 7 | 0  | 66 (80.5) |
| Ventricular Shunt | 51 | 0 | 0  | 51 (62.2) |
| ETV    | 8  | 0 | 0  | 8 (9.8)   |

*Groups are detailed in “Patients and Methods,” CM II: Chiari malformation Type II; ETV: Endoscopic third ventriculostomy
with SONTD had ventriculomegaly. Dysmorphic ventricles were seen in most patients with SONTD, the commonest of which was colpocephaly that was found in 42 patients (51.2%), which is remarkably higher when compared with the 14 cases described by Noorani et al.\[9\] in a review of 3411 consecutive cranial CT scans.

Moreover, 10-20% of children with SONTD and CM II may not develop hydrocephalus and do not require shunt placement.\[14,20\] In the present series, 19.5% (16/82) did not develop hydrocephalus during follow up period, which ranged between 1 and 12 years in Group “C”.

The aetiology of hydrocephalus in SONTD is not known with certainty. Although there are several theories, it has been demonstrated that 80-90% of children born with MMC are affected with CM II, aqueductal stenosis or fourth ventricular outflow obstruction causing non-communicating hydrocephalus, and in some cases under-development of arachnoid granulations may result in communicating hydrocephalus.\[8\] In the present series, CM II was seen in 86.6% (71/82) of patients with SONTD. It caused non-communicating hydrocephalus due to small, crowded posterior fossa in 96.6% (57/59) of Group “A”. In the remaining two patients of Group “A” hydrocephalus was caused by isolated aqueductal stenosis.

In a small proportion of patients, changes associated with CM II may not be severe enough to cause active hydrocephalus. In our series although patients in Group “B” had ventriculomegaly, they remained compensated, and did not develop active hydrocephalus during their follow up. On the other hand, 7 of 16 patients in Group “C” had CM II without hydrocephalus. It has been observed in all groups that patients did not develop active hydrocephalus within 6 months after birth remained free of hydrocephalus even if they have ventriculomegaly.

Our unit protocol when patients born with SONTD were referred for treatment was to look for hydrocephalus in the brain image before repairing the spinal defect. Twenty three patients (37.8%) in Group B and C were found to have normal size ventricles (16/23) or moderate ventriculomegaly (7/23), they did not have CSF collection or leak at the SONTD operative site, and did not need CSF diversion during the period of follow up. Patients in Group B were followed up closely in the first year of life with serial cranial scans, they continued to develop near normally and parents were satisfied by their progress. Although spina bifida team was alerted about

![Figure 6](image1.png)

**Figure 6:** Sagittal T2WI of the lumbar spine for a child, soon after birth, showing a large myelomeningocele, and low lying cord (a). There was evidence of CM II in the craniocervical junction cuts (b). A cranial MRI showed normal ventricles (not shown). The patient did not develop hydrocephalus as shown in MRI obtained after 2 years (c)

![Figure 7](image2.png)

**Figure 7:** Axial T2WI for a child with SONTD, CM II and shunted hydrocephalus, showing a large intra ventricular cyst (a). The cyst was treated by endoscopic fenestration and re-adjustment of the ventricular catheter. Note the slight change in cyst size (b). Axial T2WI taken 2 years (c) and 3 years (d) later, showing collapse of the cyst and decrease in the size of the lateral ventricles.
this group of patients, they remained symptom free and did not develop intracranial hypertension secondary to hydrocephalus.

Seventy two per cent of patients with SONTD (59/82) required treatment of active hydrocephalus, whether at the same time or after repair of the MMC. Fifty one patients had ventricular shunts and eight patients were treated with ETV. The risk of shunt failure appeared to be the same whether inserted during MMC repair or within one week from the repair, but this was significantly reduced if the ventricular shunt was inserted after one week of MMC repair. On the other hand the risk of shunt infection is significantly increased if shunt placement and MMC repair are delayed for more than 32 days after birth.[13]

After the introduction of shunting, the outcome of children with SONTD improved. Most of the deaths occur in the first year of life, mostly due to renal and respiratory problems associated with spina bifida. Only a few of the deaths are related to hydrocephalus. In a review of children treated in 1980s 27% had died, most of them in the first year of life, from causes not related to hydrocephalus but to spina bifida.[14] In the present series, 2 patients died due to respiratory failure and complications related to hydrocephalus and shunts.

The life expectancy of children with MMC has improved but it should be born in mind that modern intensive management of these patients has been pursued only for the last 45 years.[15] Of interest is that a study of long-term outcome noticed a higher mortality rate among shunted children with hydrocephalus and MMC than among non-shunted children.[21] The difference was assumed to be due to shunt related complications, and the authors suggested that efforts should be directed towards delaying shunting as much as possible or considering ETV as an alternative. There continues to be a small risk of mortality for these patients even throughout adulthood. In a recent survey of adults with spina bifida, 6% of patients died due to shunt related problems or after craniovertebral decompression for Chiari malformation.[7,13]

CONCLUSIONS

Hydrocephalus is commonly seen in patients with SONTD having CM II, which is primarily caused by congenital posterior fossa hypoplasia sufficient to cause obstruction to CSF flow, and patients with SONTD-associated hydrocephalus are likely to remain shunt dependant.

It may be advisable to delay treatment in absence of active hydrocephalus as patients with normal size ventricles or moderate ventriculomegaly may not develop hydrocephalus.

The multidisciplinary management of patients with SONTD has resulted in significant improvement in mortality and functional morbidity, achieved through regular follow up and timely interventions. Endoscopic treatment was found to be useful, but in infants with CM II will need a multicentre randomized trial to be assessed.

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