Surgical management of open spinal dysraphism: 5 years experience in Upper Egypt - a multi-centre study

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ABSTRACT

Objective: to review the surgical outcome of 60 patients with open spinal dysraphism managed in Upper Egypt, over a period of 5 years.

Patients & Methods: This retrospective study includes patients managed for open spinal dysraphism over a period of 5 years (January 2010 - December 2014) in Departments of Neurosurgery at Sohag University Hospital, Qena University Hospital, and Assiut University Hospital. We reviewed 60 patients regarding their clinical presentation, site and type of lesion, associated congenital anomalies, surgical technique, outcome, and complications.

Results: Sixty patients with open spinal dysraphism were included; thirty-seven patients (61.7%) were males while twenty-three patients (38.3%) were females. Most patients were presented at birth, or shortly after (Fifty-eight), one patient at three months, and one at 1 year. Forty-eight patients had Myelomeningocele (80%), and 12 patients had meningocele (20%). Most common level was Lumbo-sacral in Forty-six cases (76.7%), followed by Lumbar region in nine cases (15%), dorsal in three cases (5%), and two cervical cases (3.3%). Regarding clinical presentation, thirty-eight patients presented with paraplegia (63.4%), eleven patients had weak feet (18.3%), and eleven patients had good leg movements (18.3%). Three patients presented with ruptured sac. In fifty-five patients, primary closure was possible (91.7%), and rotational skin flap was needed for closure in 5 cases (8.3%). Hydrocephalus was present in twenty-seven patients (45%); nineteen patients with concurrent Hydrocephalus (31.7%), and eight patients with post-repair Hydrocephalus (13.3%). Associated congenital anomalies included: equine varus in eighteen patients (30%), kypho-scoliosis in fifteen patients (25%), cardiac anomalies in three patients (5%), and hypospadias in two patients (3.3%). Reported complications were: CSF collection in nine patients (15%), CSF leak in five cases (8.3%), Urine retention and/or abdominal distention in 3 cases (5%), Wound infection and dehiscence in 2 patients (3.3%), and one case of mortality from sleep apnea (1.6%). With follow up, secondary tethered cord occurred in fourteen patients (23.3%), Symptomatic chiari malformation and syringomyelia in three patients (5%), and dermoid cyst in one patient (1.6%).

Conclusion: Spinal dysraphism is still a challenging health problem, especially in developing countries with poor socioeconomic status. Its management requires a multidisciplinary team approach. Aim of surgical management was to prevent further deterioration, control of hydrocephalus or leak from ruptured sac.

Key words: Spinal dysraphism, Spina bifida.

Introduction:

Spinal dysraphism (Spina bifida) involves a spectrum of congenital anomalies represented as a defective neural arch through which meninges or neural elements are herniated, leading to a variety of clinical manifestations. They comprised of open and occult types. Spina bifida manifesta or (open type) is usually when there is a visible lesion associated with skin defect with an impending risk of CSF leak. In Spina bifida occults (occult type), there is normal skin coverings. Meningocele, myelomeningocele, lipomeningomyelocele, myeloschisis and rachischisis are the usual names associated depending on the pathological findings. Meningocele involves only the meninges with no neural involvement; others have variable extent of neural involvement. The majority of cases are classified as Myelomeningocele, in which the spinal cord herniats through the spinal column, resulting in nerve damage and physical disabilities including lower limb paralysis and distured bladder or bowel function. [1]

The overall management of a child with open spinal dysraphism is usually complex and prolonged. Mortality rates of 65–75% have been found in untreated patients in the first 6 months, and mortality rates of 30–40% in the first year and 50–60% in the first 3–5 years. [2]

Spinal dysraphism affect an average of 1 in every 1000 established pregnancies world wide [3]. The causes remain uncertain, with both environmental and genetic factors playing roles. Prenatal maternal factors such as exposure to alcohol, valproic acid, carbamazepine, or isotretinoin; hyperthermia; malnutrition (especially folic acid deficiency); diabetes; and obesity all increase the risk of giving birth to a child with Spina bifida.[4]

There is strong evidence that there has been a decline in incidence in developed countries since the 1970s. This is probably due to dietary fortification and advanced prenatal diagnosis with more elective terminations. Prenatal diagnosis involves alpha-fetoprotein screening and ultrasonography [5].

In developing countries like Egypt, and especially in Upper Egypt area, the incidence and prevalence of spinal dysraphism is still high due to poor socioeconomic status of the patients, poor antenatal care, consanguineous marriages, and religious factors that prevent elective terminations even after antenatal diagnosis [6]. Also, patients at presentation are usually in sepsis and malnutrition, which makes early surgical intervention difficult with more complications.

Open spina bifida is compatible with postnatal survival, although the resulting neurological impairment below the level of the lesion can lead to lack of sensation, inability to walk and incontinence. Also it may be associated with hydrocephalus, vertebral deformities, chiari II malformations, genitourinary and gastrointestinal disorders.[3]

Chiari II malformation occurs in over 90% of people with myelomeningocele. The other CNS factor that affects cognitive functions, behavior, and adaptation is hydrocephalus, and its treatment. Some form of ventriculomegaly is usually present in people with myelomeningocele because of the obstruction caused by the Chiari II malformation, and in some people with myelomeningocele or other spinal dysraphisms, because of aqueductal stenosis. Shunt diversion, which began in the 1970s, has been credited with increasing the survival rate of people with myelomeningocele. It was routine to repair the spinal lesion and implant a shunt for myelomeningocele. However, because of concerns about the long-term effects of shunt diversion due to malfunction and infection, many centers now implant shunts at birth only when there is significant ventricular dilation and monitor ventricular dilation over time with serial neuroimaging.[7]

Treatment for spina bifida includes surgery and physiotherapy. Surgery to close the newborn's spinal opening is generally performed within 24 h after birth to minimize the risk of infection and to preserve existing function in the spinal cord. However, regular monitoring, ongoing therapy, and medical and/or surgical treatments are often necessary to prevent and manage complications throughout the individual's life. Although many advances have been made in the treatment of spina bifida, resulting in increased life expectancy and improved quality of life for individuals with the disease, no treatment exists that will completely eliminate the serious disability or premature mortality associated with it. For these reasons, reducing the risk of spinal dysraphism is an important goal. [8]

Surgical repair *in utero* for early open spina bifida has been practised in several centres in the USA for the past 15 years. In The Management of Myelomeningocele Study (MOMS) [9], the trial showed that fetal surgery brings significant short-term benefits for the newborn child, including a 50% reduction in shunting for hydrocephalus and a significant improvement in spinal neurological function. Against this was a significantly higher rate of premature birth and maternal complications such as uterine dehiscence at the operation site in the *in utero* group. [3]

The aim of our study is to review the surgical outcome of 60 patients with open spinal dysraphism managed in Upper Egypt (3 main cities in Qena, Sohage and Assiut) over a period of 5 years.

Patients and Methods:

This study is a prospective study that was conducted over a period of 5 years (from January 2010 - December 2014), in the department of Neurosurgery, at 3 major centers in Upper Egypt (Qena University Hospital - Qena, Sohage University Hospital - Sohage, and Assiut University Hospital - Assiut). It included 60 patients with open spinal dysraphism.

Inclusion criteria: patients with open spinal dysraphism (spina bifida aperta) that were presented to our out patient clinics and were treated with surgical repair during the study period, and were followed up for at least 2 years.

Exclusion criteria: spina bifida occulta, and previously operated cases were excluded

from the study. We also excluded patients who failed to be followed up for at least two years post operatively.

After history and examinations, CT-Brain was done for all patients. We collected patients' clinical presentation (sex distribution, age at presentation, leg movements, sac condition, level and type of the lesion), surgical technique, associated anomalies, and complications. We followed patients in our out patient clinics, and they were referred to physiotherapy and orthopedics departments for further management. With follow up for a period varied between 2 to 5 years, late sequelae were recorded.

Surgical technique:

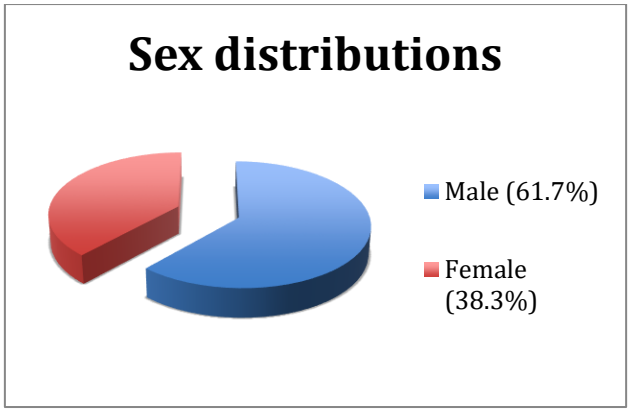
All patients were operated under general anesthesia, in prone position, and surgical technique comprised of: excision of the sac while preserving the neural tissues, then reinsertion of neural tissues into the spinal canal, with reconstruction of the dural sac, then closure of the fascial layer, and finally skin & subcutaneous tissue were closed primarily in most cases with no drain. In cases with large defect, rotational skin flap was done.

Results:

Sex distribution and Age at presentation:

- The study involved 60 patients, verified into 37 males (61.7%) and 23 females (38.3%) with open spinal dysraphism. See figure 1.
- 58 patients (96.6%) presented at birth or shortly after, one patient (1.7%) presented at 3 months' age, and one patient (1.7%) at 1 year.

Figure 1: sex distributions



Type and Level of the lesions:

- After examining the lesions of our patients we found that:
- 48 patients (80%) had Myelomeningocele, while 12 patients (20%) had Meningocele. see Table 1
- According to the level: 46 cases were in the lumbosacral area (76.7%), 9 cases in lumbar region (15%), 3 cases in dorsal area (5%) and 2 lesions in the cervical region (3.3%). See Table 2

Table (1): Type of lesions found:

Lesion	Number of patients	Percentage
Myelomeningocele	48	80%
Meningocele	12	20%

Table (2): Level of the lesion:

Site of the lesion	Number of patients	Percentage
Lumbosacral	46	76.7%
Lumbar	9	15%
Dorsal	3	5%
Cervical	2	3.3%

Clinical presentations:

- We evaluated patients regarding their clinical presentations into 3 categories: complete paraplegia, weak leg movements, and good leg movements.
- 38 patients were paraplegic (63.4%), 11 patients had weak leg movements (18.3%), and 11 patients with good leg movements (18.3%). See Table (3)
- We also categorized patients into those presented with ruptured sac and CSF leak (3 cases) and those with intact sac (57 cases). See Table (4)

Table (3): Motor weakness

Motor Power	Number of patients	Percentage
Paraplegic	38	63.4%
Weak Leg movements	11	18.3%
Good Leg movements	11	18.3%

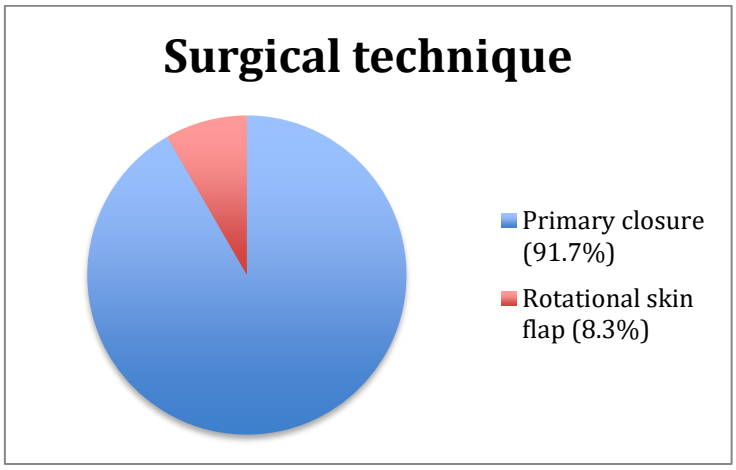
Table (4): Sac integrity

Sac Integrity	Myelomeningocele	Meningocele
Ruptured (3 cases)	2	1
Intact (57 cases)	46	11

Surgical technique:

- We divided patients into 2 groups depending on whether primary surgical closure was done or rotational skin flap was needed due to a large defect.
- Primary closure was done in 55 patients (91.7%), and rotational skin flap was used in the remaining 5 cases (8.3%). See Figure 2

Figure (2): Surgical technique:



Associated congenital anomalies (Table 5):

- Associated congenital anomalies were recorded as in the following table:

Table (5): Associated congenital anomalies

Congenital Anomaly	Number of patients	Percentage
Hydrocephalus	27	45%
Concurrent Hydrocephalus	19	31.7%
Post repair Hydrocephalus	8	13.3%
Equine varus	18	30%
Kyphoscoliosis	15	25%
Cardiac anomalies	3	5%
Hypospadias	2	3.3%

Complications & Late sequelae:

- Complications were recorded and summarized in the following table 6:

Table (6): Complications

Complications	Number of patients
CSF collection	9 (15%)
CSF leak	5 (8.3%)
Urine retention and/or abdominal distension	3 (5%)
Wound infection and dehiscence	2 (3.3%)
Mortality*	1 (1.6%)

* Cause of mortality: sleep apnea

After follow up for at least 2 years up to 5 years, the following late sequelae were recorded in the following table 7:

Table (7): Late sequelae

Late sequels	Number of patients
Tethered cord	14 (23.3%)
Symptomatic chiari and syringomyelia	3 (5%)
Dermoid cyst	1 (1.6%)

Discussion:

Our study included 60 patients, who were presented with open spinal dysraphism and managed at our university hospitals during the study period. The male (61.7%) to female (38.3%) ratio was nearly 2:1. Comparing to similar studies in other developing countries with similar socioeconomic status, this was similar to a study in Nigeria done by Idowu et al. in which he studied 36 patients with open spinal dysraphism and showed male (24) to female (12) ration of 2:1 as well.[2] In a study by Banskota et al. in Nepal, 41 cases were studied, and they showed male (58.5%) to female (41.5%) ratio of 1.4:1.[10] Another study in India, by Kumar et al., showed male to female ratio of 1.5:1.[11]

In our study, the age at presentation was mostly at birth (96.6%), which was similar to Idowu et al., but different from Banskota et al. as the mean age of presentation was 2.71 years. In Kumar et al. he showed even higher mean age at presentation of 5.7 years.

Myelomeningocele represented 80% of patients in our study, which is similar Banskota et al. (also 80% of cases), and was higher than a study done in India by Kumar et al. In his study, 119 cases of open spina bifida were included, and Myelomeningocele represented 72% of them. [11]

The most common level in our study was Lumbosacral area with 76.7%, followed by the lumbar region (15%). This was similar to Kumar et al. (44% in Lumbosacral area, 32% in dorsolumbar area). This was different from Banskota et al., as the most common level was lumbar with 66%, followed by lumbosacral region with only 29%.

Regarding clinical presentations, 38 patients (63.4%) were paraplegic (all were Myelomeningocele), 11 patients had good leg movements (all were Meningocele), and the remaining patients (11 patients) had weak legs. Patients with weak legs included one meningocele (which was ruptured at presentation), and the rest were myelomeningoceles. All patients presented with intact sac, except 3 cases that presented with ruptured sac with CSF leak (one meningocele and two myelomeningocele). The ruptured meningocele had weak leg movement, and both ruptured myelomeningocele cases were paraplegic. In 55 patients, primary surgical closure was possible, but rotational skin flap was needed in 5 patients only.

Hydrocephalus was the most common associated congenital anomaly, which was present in 27 patients (45% of the cases). In 19 patients, the hydrocephalus was present concurrently with spina bifida, while in the remaining 8 cases, Hydrocephalus developed post repair. In all cases, ventriculoperitoneal shunting was done in all patients. Banskota et al. found in his study that Hydrocephalus was present in 51% of the cases; about half of them (24%) developed after surgical repair. Shunting was done in 75.6% of his cases. This also, was similar to Kumar et al. with 46% of his cases developed Hydrocephalus, with only 85% of them required shunting.

Other associated anomalies included, congenital talipes equinovarus (30% of the cases), kyphoscoliosis (25%), cardiac anomalies (5%), and hypospadias (3.3%). In Kumar et al., he found scoliosis in 36% of the cases, equinovarus in 33%, and chiari malformation in 45% of the cases.

We encountered CSF collection in nine patients (15% of our cases), which was resolved spontaneously with positioning. In 5 cases (8.3%), CSF leak occurred postoperatively, and all resolved after shunting due to Hydrocephalus. Urine retention and abdominal distension occurred in 3 patients (5%), which improved with conservative measures. Wound infection and dehiscence occurred in 2 patients (3.3%), and was treated by debridement and 2ry suturing. We only had one case of

postoperative mortality from sleep apnea (1.6%). There was no deterioration in neurologic condition of the patients. Compared to Kumar et al., he had CSF leak in 33% of the cases, pseudomeningocele in 17%, wound infection in 14%, meningitis in 8%, and two cases of postoperative mortality. Idowu et al. also had one case of postoperative mortality in his series of 36 patients.

The duration of follow up in our study was between 2 to 5 years. Secondary tethered cord occurred in 14 patients (23.3%), and untethering was done in 9 patients (15%). Symptomatic chiari malformation and syringomyelia were detected in 3 patients (5%), and were treated by Craniocervical decompression. We also encountered one case of dermoid cyst. It should be noted that in our study, we didn't perform preoperative MRI of the neurospinal axis routinely due to young age at presentation. So, we cannot comment on the incidence of associated chiari malformation in our series, but with follow up, only 3 symptomatic cases were encountered as mentioned above. Comparing to a study by Talamonti et al. in Italy, in which he studied the long term follow up in 202 patients with Myelomeningocele with a follow up period of 1 to 25 years, he found tethered cord in 20% of the cases, with only 11% needing surgery. In his study, he found hydrosyringomyelia in 68% of the cases.[12]

Conclusion:

Despite the decline in incidence of spinal dysraphism worldwide, it is still a challenging health problem, especially in developing countries with poor socioeconomic status. In our study of 60 patients in the area of Upper Egypt, the early surgical intervention and the young age at presentation, helped in improving outcome, and decreasing the surgical complications. The aim of surgical management was to prevent further deterioration, control of hydrocephalus or leak from ruptured sac. This requires multidisciplinary team management of Pediatrics, Neurosurgery, Orthopedics, Urology, and Rehabilitation. Follow up is mandatory for prolonged period of time, as a considerable percentage of patients develop late sequelae that need special management.

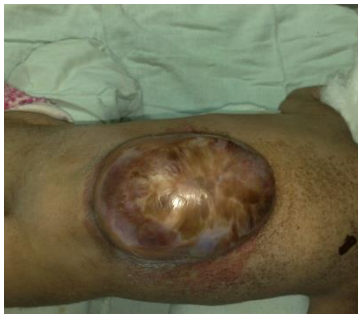
Limitations and Recommendations:

The limitations of our study includes, the short period of follow up, as more time is needed to determine long term results of the cases. Urologic complications of our patients were not evaluated due to young age of patients.

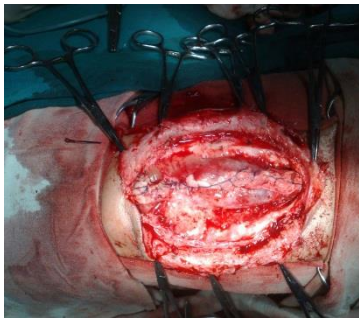
Further study is needed to determine the important risk factors in our regional areas for spinal dysraphism, and the public health programs employed to decrease the incidence of these cases.

Case presentation

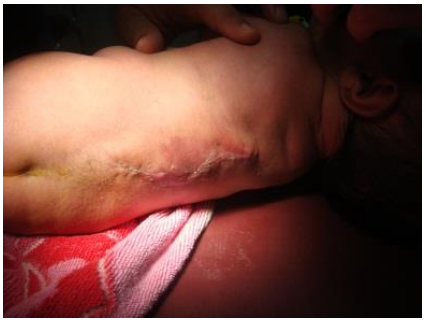
Dorsolumber myelomeningocele



Pre operative

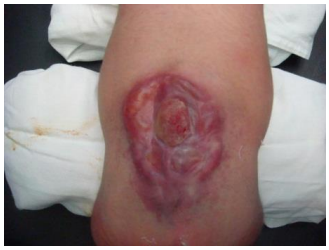


Operative photo

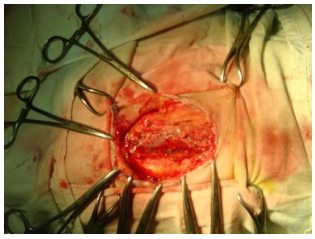


Pos operative

Lumbosacral Myelomeningocele



Pre operative



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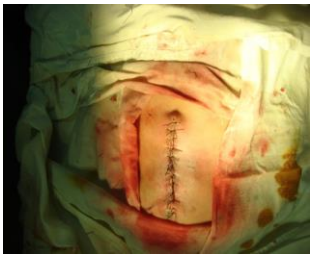
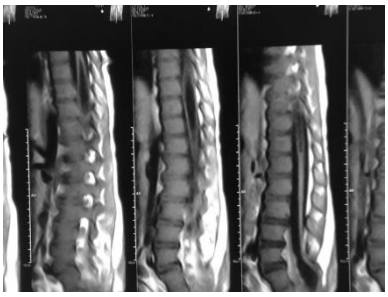
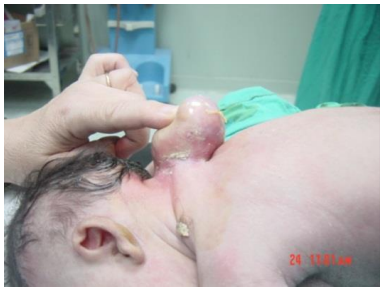


Closure



Post operative tethered cord



Cervical meninigocele

References

1. Venkataramana NK. Spinal dysraphism. **Journal of Pediatric Neurosciences**. 2011;6(Suppl1):S31-S40.
2. Idowu, O.E., Apemiye, R.A. Outcome of myelomeningocoele repair in sub-Saharan Africa: the Nigerian experience. **Acta Neurochir**. 2008;150:911–913 (discussion 913. Epub 2008 Aug 23).
3. Copp AJ, Stanier P, Greene NDE. Neural tube defects – recent advances, unsolved questions and controversies. **Lancet neurology**. 2013;12(8):799-810.
4. Liptak GS, Dosa NP. Myelomeningocele. **Pediatrics in Review**. 2010; 30(31): 443–450.
5. Netto JMB, Bastos AN, Figueiredo AA, Pérez LM. Spinal Dysraphism: A Neurosurgical Review for the Urologist. **Reviews in Urology**. 2009;11(2):71-81.
6. Salih MA, Murshid WR, Seidahmed MZ. Epidemiology, prenatal management, and prevention of neural tube defects. **Saudi Medical Journal**. 2014;35(Suppl 1):S15-S28.
7. Fletcher JM, Brei TJ. Introduction: Spina Bifida—A Multidisciplinary Perspective. **Developmental disabilities research reviews**. 2010;16(1):1-5.
8. Yi Y, Lindemann M, Colligs A, Snowball C. Economic burden of neural tube defects and impact of prevention with folic acid: a literature review. **European Journal of Pediatrics.** 2011;170(11):1391-1400.
9. Adzick NS, Thom EA, Spong CY, et al. A randomized trial of prenatal versus postnatal repair of myelomeningocele. **N Engl J Med**. 2011;364:993–1004
10. Banskota, N et al. Surgical Management of Spinal Dysraphism: Five -year Experience in a Central Hospital. **Journal of Nepal Paediatric Society**, [S.l.], v. 34, n. 1, p. 34-38, mar. 2014. ISSN 1990-7982.
11. Kumar R, Singh SN, Spinal dysraphism: trends in northern India: **Pediatr Neurosurg** 2003; 38(3):133-45.
12. Talamonti, G., D'Aliberti, G., Collice, M. Myelomeningocele: long-term neurosurgical treatment and follow-up in 202 patients. **J Neurosurg**. 2007;107:368–386.