

# Cystic Cervical Dysraphism: A Single Institution Experience and Outcome from a Case Series

MOHAMED GABR, M.D.; MOHAMED R. NAGY, M.D. and AHMED A. ABDULLAH, M.D.

*The Department of Neurosurgery, Faculty of Medicine, Cairo University*

## Abstract

**Background:** Cystic cervical dysraphism is considered a rare entity. It has more frequent congenital anomalies than their lumbosacral variety. Most of these lesions are symptomatic in childhood period due to progressive tethering of spinal cord or nerve root.

**Material and Methods:** This is a retrospective study of all cases underwent surgery for cystic cervical dysraphism from July 2016 to July 2021 at the Aburish Pediatric Hospital, Cairo University. All patients were neurologically and radiologically assessed. Neurological outcome was noted at the last follow-up.

**Results:** Twelve children (7 females and 5 males) were operated upon. The mean age was  $20.8 \pm 13.8$  (range, 4-48) months. Meningocele was found in five patients (41.7%). Other five children (41.7%) had a thick stalk with lipomatous nodular swelling. One patient (8.3%) had myelocystocele. Another patient (8.3%) was suffering from myelomeningocele. Five cases were hydrocephalic; two of them underwent ventriculoperitoneal shunt insertion prior to surgery. Associated anomalies were present in nine children (75%). For the neurological outcome, we have noted no signs of neurological deterioration after surgery in all patients.

**Conclusions:** Provided that there is no or minimal neurological deterioration at time of presentation, the outcome of surgery for cystic cervical dysraphism is excellent. The surgery helps in improving the cosmetic results, preventing the development of further neurological deterioration, and minimizing the infection. This is achieved mostly by efficient untethering.

**Key Words:** *Cervical dysraphism – Outcome – Complications – Meningocele – Myelomeningocele.*

## Introduction

**SPINAL** dysraphism refers to a group of congenital disorders that result in a weakened neural arch, allowing the meninges or other neural components to protrude, resulting in a variety of clinical symptoms [1].

Cervical spina bifida, whether occult or overt, is a very rare condition that is often linked with

other CNS and spine problems [2]. There are two types of anomalies in cervical spina bifida cystica: a myelocystocele that herniates posteriorly into a meningocele and a meningocele with or without an underlying split cord abnormality [3]. Cervical myelocystoceles (CMC) and cervical meningoceles (CM) are uncommon spinal dysraphic lesions that affect 4-8% of cases with spina bifida cystica [4,5]. Children with meningoceles typically have normal neurological development.

Tethering of the cord is the main problem with cervical dysraphism (CD). Even if asymptomatic at birth, tethering will cause progressive neurological deficits mainly presenting as upper limb weakness which may be delayed. The surgical repair of these lesions is of utmost importance to achieve better cosmesis, and intradural exploration allowing untethering of the cord and preventing delayed neurological deterioration [2,6,7]. It is difficult to follow-up the prognosis of surgical treatment of these lesions due to their relative rarity and the associated congenital anomalies increasing the mortality of affected infants [8]. However, cervical meningoceles are usually associated with more favorable clinical outcome [9].

We discussed our experiences with children with cervical dysraphism as a separate type of spinal dysraphism, pointing out the differences between them and their more prevalent lower dorsal/lumbar spinal counterparts.

## Material and Methods

Between July 2016 and July 2021, registered cases reports were revised and cases of cervical dysraphism were collected at the Aburish Pediatric Hospital, Cairo University. All records were evaluated retrospectively, and relevant data, including demographics, the size of the lesion, the type of lesion, and concomitant abnormalities, was obtained from hospital reports, outpatient records, and the digital information system.

**Correspondence to:** Dr. Ahmed A. Abdullah,  
[E-Mail: ahmedarahman@kasralainy.edu.eg](mailto:ahmedarahman@kasralainy.edu.eg)

All children underwent neurological and radiological assessment. Magnetic resonance imaging (MRI) of the brain and entire spine was done for all children. Cranial ultrasonography or computed tomography (CT) of the brain was used for screening of hydrocephalus. CT of the cervical spine was done for eight children. All cranial and spinal imaging were revised to identify any accompanying anomalies such as Chiari malformation (CM), hydrocephalus, and syringomyelia. An X-ray of the entire vertebral column was done to screen for curvature anomalies. Echocardiography was implicated in children with associated congenital cardiac anomalies.

The patients were treated surgically with sac excision and intradural sac exploration using standard microsurgical procedures. To locate the normal dura, laminectomy was conducted at least one level above and below the affected portions. Meningocele was found in 5 patients. A thick tube like stalk projecting from the posterior aspect of the spinal cord was observed to be retained in the sac in the other five individuals. Through a defect in the posterior midline structure, the stalk pierced the meningeal covering. The stalk was amputated a few millimeters from the spinal cord. A cystic growth was discovered inside the projecting dural sac in one case. The internal cyst communicated with the syringomyelic spinal cord, whereas the outside cyst communicated with the subarachnoid region (myelocystocele). Another patient had a myelomeningocele. Simple dural closure was possible for all cases without dural grafting. The procedure was uneventful in all cases. The procedure records were used to track the operational information. Cases that required redo surgery has been reviewed and indications and operative findings were reported. Surgery-related complications were also reported.

The children were followed-up regularly in the outpatient clinic monthly for the first three months then every two months later. The follow-up records were collected, and some patients were contacted by telephone. Pre- and post-operative neurological deficits were documented.

Data were analyzed and statistical significance was calculated using nonparametric tests.

### Results

The study included twelve children: Seven females (58.3%) and five males (41.7%). The mean age was  $20.8 \pm 13.8$  (range, 4-48) months.

Presentation varied from cervical swelling with no symptoms (8/12), urinary problems (3/12) (retention with overflow, incontinence in two cases, and incontinence with loss of bladder sensation in one case), and delayed walking (2/12). None of the patients had a CSF leak or dimple when they on presentation. One patient was referred to us after undergoing surgery for cervical swelling at a different center after which he experienced recent onset of walking difficulty and urinary frequency; persistent tethering was discovered during the assessment.

Meningocele was found in five patients (41.7%) (Figs. 1,2,3). Other five children (41.7%) had a thick stalk with lipomatous swelling. The stalk was found attached to the swelling on the posterior surface of the cord, emerging from a defect in the posterior osseous parts. One patient (8.3%) had myelocystocele. Another patient (8.3%) was suffering from myelomeningocele. This myelomeningocele patient also had hydrocephalus and a Chiari type II malformation.

Five cases were hydrocephalic, two of them underwent ventriculoperitoneal (VP) shunt insertion prior to surgery, and the other three were managed conservatively as they had an open head and moderate ventriculomegaly. The rest of the cases were operated by direct repair, untethering of the cord, and surgical excision.

Associated anomalies were present in nine children (75%). Split cord malformation (SCM) was seen in four of the twelve patients (25%). Surgery for SCM was required in all four patients; in three of them, a fibrous spur was excised at the same level; and in one of them, a different level of laminectomy (D7) was needed to remove the bony spur. There were 3 patients (75%) who had a syrinx.

Corpus callosum agenesis (CCA) affected three of the twelve children (25%). Tethering was found at the C2 level in all three CCA patients, and syrinx was confined to the cervical spine only in all three. Postoperative CSF diversion was needed for wound leakage in one CD patient with CCA and ventriculomegaly. Five of the patients (41.7%) had Chiari malformation. Three of the five CD with CM patients had urinary problems, while one patient had difficult walking. Two of the patients were suffering from hydrocephalus. SCM was present in one of the patients (20%). Three of the patients were suffering from syrinx.

For the neurological outcome, no patient showed signs of neurological aggravation after surgery. Three patients needed additional surgery:

two needed a VP shunt prior to surgery, and the other needed a shunt after surgery due to leaking

CSF from the surgical wound. The mean follow-up period was  $32.7 \pm 17.5$  (range, 5-66) months.

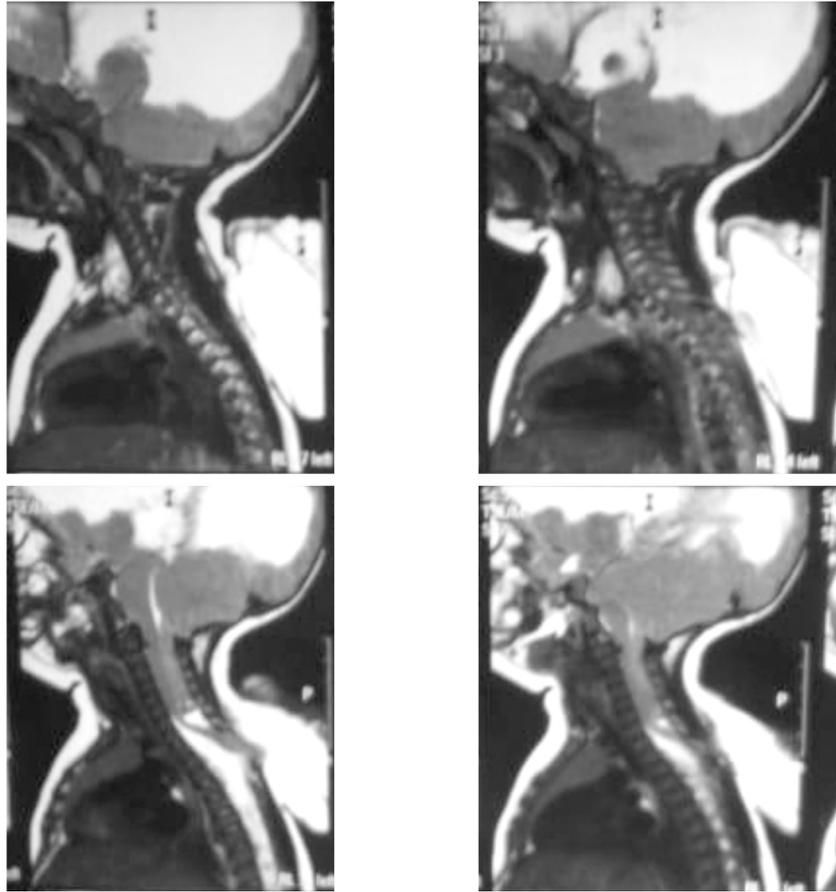


Fig. (1): The preoperative sagittal MRI cuts of the cervical spine of 4-months old boy showing a large meningocele.

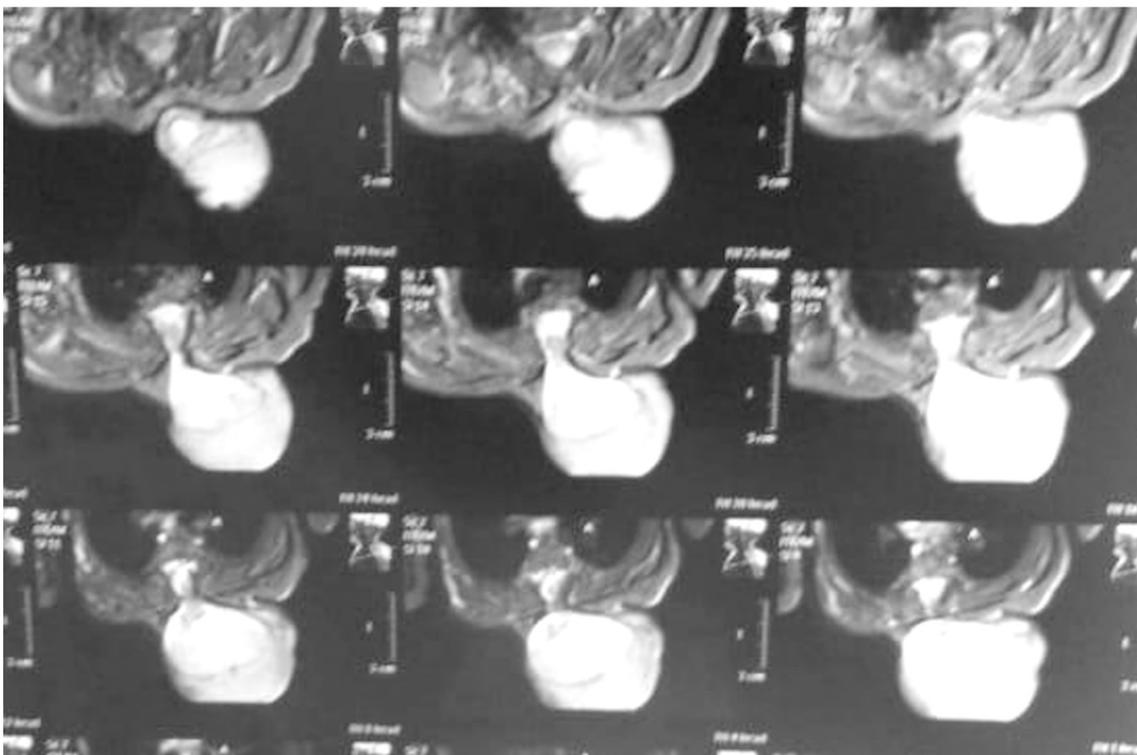


Fig. (2): The preoperative axial MRI cuts of the cervical spine of the same boy. The patient was operated by surgical excision and repair.

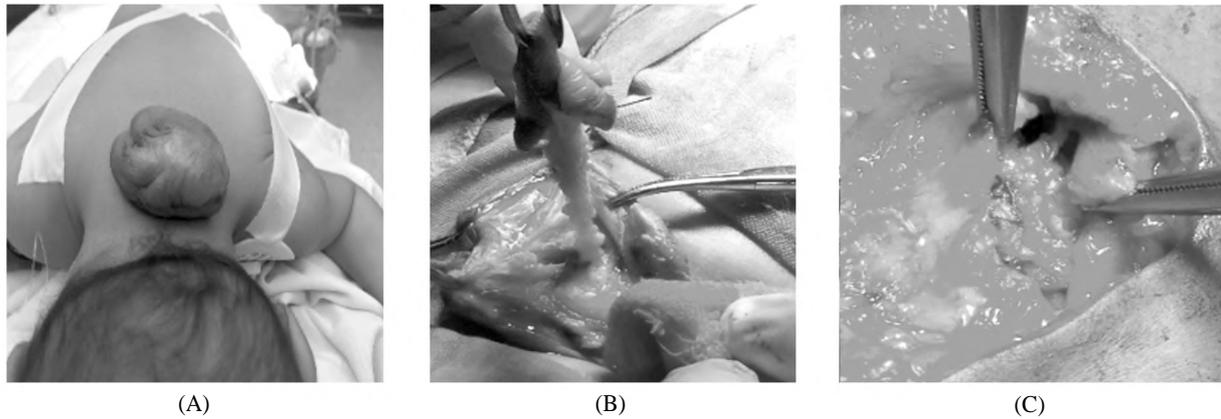


Fig. (3): Intra operative photo of the same boy showing: (A) The cervical meningocele before excision, (B) Fibrovascular band connected to cord, and (C) Suturing and closure.

### Discussion

As a distinct clinical entity, there are very few established case series on CD [10-12]. Here we report our experience of 12 cases of cystic cervical dysraphism. The incidence of cervical dysraphism is 1%-6% [8,3,13,14]. Although Habibi et al., study revealed a 56.2 percent male prevalence [15], the majority of investigations revealed a female preponderance. A female majority of 58.3 percent was also discovered in our research.

Salih et al., highlighted the etiological factors of CD. The most prevalent causes of spinal dysraphism include low socioeconomic level, insufficient folic acid intake, diabetes mellitus, obesity, vitamin B 12 insufficiency, high body temperature, valproic acid, metabolic teratogenic agents, and environmental pollution [15].

In the early 1990s, the categorization of cystic dysraphism was modified with the inclusion of the phrase "limited dorsal myeloschisis". The terms include meningoceles and meningocystocels spectrum of dysraphism. Pang et al., further classified them based on their internal structures, such as limited dorsal myeloschisis, which has a fibro-neurovascular stalk of neurons, glia, and peripheral nerves inside a dural sac, and SCM, which has two hemi cords separated by a fibrous septum that connects the hemi cords to the dura near the limited dorsal myeloschisis stalk [6]. Salomão et al., and Habibi et al., divided them similarly [17,18].

The majority of the authors agree that CD occurs as a result of incomplete neurulation in the embryo. But for the final fusion of the apposed neural folds, much of the neurulation is thought to have occurred normally. In the dorsal midline, a thin region remains open. Despite proper neurulation, the cutaneous and neural ectoderms never

divid at this time [19]. The myofascial tissues produce a dorsal median stalk of central nervous system tissue that acts as the first link between the virtually closed neural tube and the gaping cutaneous ectoderm [2].

The minimal and delayed neurulation abnormality hypothesis is supported by the higher occurrence of normal motor activity in CD. Kasliwal et al. pointed out that CD varies from patients with dysraphism in other areas because the tethering is caused by a neuroglial stalk that emerges from the dorsal surface of the cervical cord, rather than a misplaced neural placode [21].

Of the ten cases in their series, Kasliwal et al., reported two cases of thoracico-lumbar myelomeningocele (MMC) [21]. In the study by Habibi et al., there were two cases (2/16) of lumbosacral LPMC [18]. No cases in our series had double LPMC.

Other studies have reported varying incidences of associated anomalies and three out of ten patients had hydrocephalus [23]. Salomao et al., found a 7/18 association [17], Steinbok found a 5/8 association [2], and Habibi found a 50% association [18]. Our study's incidence matches that of previous studies in the literature [18,22]. Because of the high prevalence of hydrocephalus, all CD patients should be screened. The presence of hydrocephalus alone should not be regarded as an indication of CSF diversion in the preoperative period; however, these patients should be closely monitored in the postoperative period with a low threshold of diversion.

CD appears to be exempt from the universal law of relationship between CM and spinal dysraphism. In our study, 5/12 (41.6%) of the patients had CM. In their study, Habibi et al., discovered

only a 25% association [18]. Previous studies found a higher correlation between Chiari type II malformations and the myelocystocele subgroup of CD. In our series, one patient of myelocystocele was not associated with CM [26,27].

The surgical indication for CD, unlike the thoraco-lumbar type, is essentially aesthetic or for the avoidance of motor impairments. Sac excision and detethering are required for surgical therapy of CD. In addition to sac resection, all our patients underwent intradural exploration and detethering. During the postoperative period, one patient required CSF diversion. In the study by Kasliwal et al., three out of ten patients had CSF leaks, with one requiring theco-peritoneal shunt and two requiring dural repair [21].

These individuals require a meticulous sac dissection as well as the diagnosis of the tethering's source. Because there is no placode in these circumstances, it is difficult to determine stalk attachment. A surgical exposure that includes two vertebral levels above and two vertebral levels below is indicated for adequate tubular stalk access. Even if the intraoperative and radiological examinations do not reveal tethering, the dura should be opened and the source of tethering explored. We detected an adhesion or a stalk situated dorsally in every subject in our study. The stalk should be severed at the base, and intradural exploration should take place. In these lesions, the posterior surface of the cervical spinal cord is sometimes attached to the wall of the meningocele with filamentous attachments.

Tethering's specific process is unknown; nevertheless, in their investigation, Duz et al., demonstrated how band may lead to tethering [28]. The process was assumed to be similar to that of compressive cervical spondylotic myelopathy. The dorsal traction force of the tethering stalk or adhesion might be the source of the likely neurological degeneration induced by the cervical spinal cord stretch injury.

#### *Conclusion:*

Cervical spine dysraphism is an uncommon disorder that differs from lumbosacral dysraphism in terms of clinical manifestations. CD patients are less likely to have CSF leaks or motor weakness, making them safer to operate on. The two most essential reasons to operate are early detethering and cosmetic indicators. Patients with CD should be checked for any abnormalities that may be present.

#### *Declaration of patient consent:*

All appropriate patient permission documents were received, according to the authors. By signing the consent form, the patient(s) has/have given his/her/their permission to have his/her/their images and other clinical information published in the journal. The patients are aware that their first and last names will not be published, and that while every effort will be taken to keep their identities hidden, anonymity cannot be guaranteed.

#### *Financial support and sponsorship:*

Nil.

#### *Conflicts of interest:*

There are no conflicts of interest.

### **References**

- 1- VENKATARAMANA N.K.: Spinal dysraphism [Internet]. Vol. 6, Journal of Pediatric Neurosciences. Wolters Kluwer Medknow Publications, [cited 2021 May 1]. p. S31. Available from: /pmc/articles/PMC3208922/, 2011.
- 2- STEINBOK P.: Dysraphic lesions of the cervical spinal cord. Neurosurgery Clinics of North America, 6 (2): 367-76, 1995.
- 3- VENKATARAMANA N.K.: Spinal dysraphism. Journal of Pediatric Neurosciences, 6 (Suppl. 1): S31, 2011.
- 4- BARSON A.J.: Spina Bifida: The Significance of the Level and Extent of the Defect to the Morphogenesis. Developmental Medicine & Child Neurology, 12 (2): 129-44, 1970.
- 5- DORAN P.A. and GUTHKELCH A.N.: Studies in spina bifida cystica: I general survey and reassessment of the problem. Journal of Neurology, Neurosurgery & Psychiatry, Nov. 1; 24 (4): 331-45, 1961.
- 6- PANG D. and DIAS M.S.: Cervical Myelomeningoceles. Neurosurgery, Sep. 33 (3): 363-73, 1993.
- 7- STEINBOK P. and COCHRANE D.D.: The nature of congenital posterior cervical or cervicothoracic midline cutaneous mass lesions: Report of eight cases. Journal of Neurosurgery, 75 (2): 206-12, 1991.
- 8- SHURTLEFF D.B. and LEMIRE R.J.: Epidemiology, etiologic factors, and prenatal diagnosis of open spinal dysraphism. Neurosurgery Clinics of North America, 6 (2): 183-93, 1995.
- 9- KUMAR R.: Comparative study of complex spina bifida and split cord malformation. Indian Journal of Pediatrics, May 72 (2): 109-15, 2005.
- 10- GUNJAN G., SHARMA S., JAIN S.K. and CHOPRA S.: Asymptomatic posterior cervical myelomeningocele with tethered cord in an adolescent: A rare form of spinal dysraphism with rare presentation. Romanian Neurosurgery, 113-7, 2016.
- 11- ARMAS-MELIÁN K., IGLESIAS S., ROS B., MARTÍNEZ-LEÓN M.I. and ARRÁEZ M.Á.: Cervical myelomeningocele with CSF leakage: A case-based review. Child's Nervous System, 1-6, 2020.

- 12- ROSENBLUM J.S., CAPPADONA A.J., ARGERSINGER D.P., PANG Y., WANG H., NAZARI M.A., et al.: Neuraxial dysraphism in EPAS1-associated syndrome due to improper mesenchymal transition. *Neurology Genetics*, 6 (3), 2020.
- 13- MUGARAB SAMEDI V., SCOTLAND J. and CLARK D.: Cervical myelocystocele: rare presentation of spinal dysraphism. *Oxford Medical Case Reports*, (4): omy007, 2018.
- 14- VALEUR N.S., IYER R.S. and ISHAK G.E.: Cervicothoracic cystic dysraphism. *Pediatric Radiology*, 46 (10): 1471-81, 2016.
- 15- SALIH M.A.: Neural tube defects: Challenging, yet preventable. *Saudi Medical Journal*, 35 (Suppl. 1): S3, 2014.
- 16- SALOMÃO J.F., CAVALHEIRO S., MATUSHITA H., LEIBINGER R.D., BELLAS A.R., VANAZZI E., et al.: Cystic spinal dysraphism of the cervical and upper thoracic region. *Child's Nervous System*, Mar. 22 (3): 234-42, 2006.
- 17- SALOMÃO J.F., CAVALHEIRO S., MATUSHITA H., LEIBINGER R.D., BELLAS A.R., VANAZZI E., et al.: Cystic spinal dysraphism of the cervical and upper thoracic region. *Child's Nervous System*, 22 (3): 234-42, 2006.
- 18- HABIBI Z., NEJAT F., TAJIK P., KAZMI S.S. and KAJBAFZADEH A.M.: Cervical myelomeningocele. *Neurosurgery*, 58 (6): 1168-74, 2006.
- 19- DIAS M.S. and PANG D.: Split cord malformations. *Neurosurgery clinics of North America*, 6 (2): 339-58, 1995.
- 20- KASLIWAL M.K., DWARAKANATH S. and MAHAP-ATRA A.K.: Cervical meningocele - An institutional experience. *Child's Nervous System* [Internet]. 2007 Nov. [cited 2021 May 15]; 23 (11): 1291-3. Available from: <https://pubmed.ncbi.nlm.nih.gov/17628809/>.
- 21- KASLIWAL M.K., DWARAKANATH S. and MAHAP-ATRA A.K.: Cervical meningocele - An institutional experience. *Child's Nervous System*, 23 (11): 1291-3, 2007.
- 22- MEHROTRA A., SINGH S., GUPTA S., PANDEY S., SARDHARA J., DAS K.K., et al.: Cervicothoracic Spinal Dysraphism: Unravelling the Pandora's Box. *Journal of pediatric neurosciences* [Internet]. 2019 Oct 1 [cited 2021 May 15]; 14 (4): 203-10. Available from: <http://www.ncbi.nlm.nih.gov/pubmed/31908661>.
- 23- MEHROTRA A., SINGH S., GUPTA S., PANDEY S., SARDHARA J., DAS K.K., et al.: Cervicothoracic spinal dysraphism: Unravelling the Pandora's box. *Journal of Pediatric Neurosciences*, 14 (4): 203, 2019.
- 24- SINGH S., MEHROTRA A., PANDEY S., GUPTA S., BHAISSORA K.S., GAJBHIYE S., et al.: Cystic Cervical Dysraphism: Experience of 12 Cases. *Journal of pediatric neurosciences* [Internet]. 2018 [cited 2021 May 15]; 13 (1): 39-45. Available from: <http://www.ncbi.nlm.nih.gov/pubmed/29899770>.
- 25- TRACY P.T. and HANIGAN W.C.: Spinal Dysraphism: Use of Magnetic Resonance Imaging in Evaluation. *Clinical Pediatrics* [Internet]. 1990 Jul 2 [cited 2021 Jun 26]; (4): 228-33. Available from: <https://journals.sagepub.com/doi/abs/10.1177/000992289002900405>
- 26- HABIBI Z., NEJAT F., TAJIK P., KAZMI S.S. and KAJBAFZADEH A.M.: Cervical myelomeningocele. *Neurosurgery*, Jun. 58 (6): 1168-74, 2006.
- 27- SUN J.C.L., STEINBOK P. and COCHRANE D.D.: Cervical myelocystoceles and meningoceles: Long-term follow-up. *Pediatric Neurosurgery* [Internet]. 2000 [cited 2021 May 15]; 33 (3): 118-22. Available from: <https://pubmed.ncbi.nlm.nih.gov/11096358/>
- 28- DUZ B., ARSLAN E. and GÖNÜL E.: Cervical congenital midline meningoceles in adults. *Neurosurgery*, 63 (5): 938-45, 2008.

## تشوهات الكيس النخاعي بالفقرات العنقية؛ خبرة المستشفى واحدة لنتائج التدخل الجراحي للحالات السريرية

الكيس السحائي للأطفال يعتبر من التشوهات الجنينية وتكون في الفقرات العنقية أو بالفقرات القطنية والعجزية.

الدراسة شملت دراسة لـ ١٢ طفل (٧ أنثى و ٥ ذكور) وأجريت الجراحة في مستشفى الأطفال بمستشفى أبو الريش الجامعي في الفترة بين (يوليو ٢٠١٦ - يوليو ٢٠٢١) حيث كان متوسط العمرى للأطفال (٤-٤٨ شهر).

تم تقييم الحالات بالفحص العصبي لهم والاشعاع للكيس النخاعي. وكانت فترة المتابعة تتراوح ما بين (٥-٦٦ شهر). الجراحة كانت باستئصال الكيس السحائي وتم تركيب صمام مخي بريتنوني في أطفال وذلك لوجود استسقاء بالمخ. تتمثل المشكلة الرئيسية في الجراحة هي التصاق العصبى للنخاع الشوكي واتصاله بالجلد الخارجى للكيس.

هناك أعراض مصاحبة مع الكيس مثل عدم التحكم فى البول وضعف فى الطرفين السفليين والهدف من الجراحة هو تصليح الشكل الخارجى باستئصال الكيس وتسليك الأعصاب الملاصقة للكيس والجلد ومتابعة ظهور أى استسقاء أو تسرب للسائل النخاعي من الجلد.

تم حدوث تدهور فى الوظائف الحركية فى ٣ من المرضى أثناء المتابعة بعد الجراحة وتم إجراء مراجعة للجراحة وذلك لتسليك الكيس النخاعي.

تم تركيب صمام مخي بريتنوني فى عدد ٢ مرضى قبل الجراحة بينما باقى المرضى تم تركيب الصمام بعد الجراحة.