Endoscopic third ventriculostomy in children below 2 years

Authors: Ahmed Saro[1], Abdin Khirallah[2]

Neurosurgery department - Sohag faculty of medicine

Abstract

Introduction:

For many years, the VP shunt was the main treatment of hydrocephalus till the recent advances in the neuroendoscopic applications. The use of the endoscopic third ventriculostomy provides an ideal treatment without complications of the traditional shunts. However, it can be challenging in the age group below 2 years as the results are unexpected and different from the adult group. Our aim in this study is to assess the results of the ETV in children below 2 years; we analyzed the children data and its relation to the outcome. We also, reported the complications happened during and after the procedure.

Patients & methods:

A retrospective and prospective study was carried out on twenty-one children who had hydrocephalus. An ETV was done for them using the (Karl Storz) neuroendoscopy between April 2012 and September 2014. All of them had an aqueductal stenosis. Follow- up CT was done for all children, one month and one year after the procedure.

Results:

Their age ranged from 6 months to just below two years, with the mean age was 14.5 ± 5.2 months. They were 15 males and six females. The overall success rate was 66.6% with the mean follow-up period of 18 months. VP shunt was inserted in 5 babies due to the ETV failure, one case of CSF leak and one case died due to intracranial hemorrhage.

Conclusion:

Our study showed the effectiveness of ETV in children less than two years with a primary aqueductal stenosis in general with reasonable complications. It is safe and effective method for treating hydrocephalus in this age group without confrontation of VP shunt and its hazards. ETV scoring system (ETVSS) is beneficial in predicting the ETV success rate.

Keywords:

ETV (endoscopic third ventriculostomy) – VP (Ventriculoperitoneal) shunt – CSF (cerebrospinal fluid) – IVH (intraventricular hemorrhage) – HC (hydrocephalus) – CNS (central nervous system) – computed tomography (CT).

Introduction:

ETV was first applied in infants. Mixter performed the first successful endoscopic ventriculostomy in a 9-months-old baby [1]. Guiot performed a successful ETV in newborn with follow up for 7 years without complications [2]. Since the 1960s the classic treatment of hydrocephalus was the shunts with its all types including programmable or non-programmable shunts. Although advances occurred in shunting system, the need for shunt revision is still high, beside the well-known complications of the shunts as infection and patient dependence [3]. Since the invention of the neuro endoscope in the last two decades, ETV became the recent acceptable option for treatment of hydrocephalus [4]. ETV is a safe, simple technique and provides the CSF shunting in a physiological pathway to the subarachnoid space [5].

However, the controversy among authors was about the success rate of ETV in children especially those below two years with the need to repeat the ETV or inserting shunt after ETV failure [6]. Some authors don’t recommend ETV below two years as it has a higher failure rate, while others reported no difference in the outcome in babies and children if compared to older patients [7]. On the other hand, some authors correlated the failure of ETV with the cause of hydrocephalus rather than the age of the patient, i.e. post hemorrhagic and postinfectious hydrocephalus have a higher failure rate of ETV. ETVSS is strongly recommended to predict the rate of ETV success [8-10].

Our aim in this study is to add data on this controversial issue and report our experience in the success rate of ETV in children with hydrocephalus secondary to a primary aqueductal stenosis below two years and report the postoperative complications like CSF leak, obstruction of the stoma and infection happened among those children.

Patients and method:

Our study was a prospective and retrospective study applied for twenty-one children between April 2012 and September 2014 diagnosed with hydrocephalus secondary to a primary aqueductal stenosis. Fifteen males and six females were included with age varying from 6 months to two years. Out of the 21 children, six children were premature. ETV was done for all babies through a burr hole just anterior to the coronal suture in the mid-pupillary line (Kocher point) using rigid endoscope (Karl Storz) with lens 0 degree. A ventriculostomy was done in the floor of the third ventricle between infundibulum and the mammillary bodies using the endoscopy forceps and widening of the opening using Fogarty catheter. A ringer lactate solution was used if there was any occasional bleeding.

Ensuring enough ventriculostomy was assured by free CSF circulation and good pulsation of the remnants of arachnoid membranes. All children underwent preoperative MRI to diagnose the cause of hydrocephalus. A postoperative CT was done for all patients after one month and a second CT follow- up was done after one year to compare the reduction in the width of the lateral ventricle. The failure of the procedure included the deterioration in child clinical condition as enlarging the head size and the radiological increase in the ventricular width and periventricular edema. The mean follow up period was 18 months.

Inclusion criteria:

1. Children below two years.
2. Hydrocephalus secondary to a primary aqueductal stenosis.

Exclusion criteria:

1. Age above two years.
2. Posthemorrhagic or post infection hydrocephalus.
3. Other CNS congenital anomaly.
4. Secondary hydrocephalus to brain tumors.

Results:

The total number was 21, fifteen males (71.4%) and six females (28.6%). The age varied from 6 months to 2 years with the mean age 14.5 ± 5.2 months. They were six babies below one year and fifteen babies from 1 to 2 years. They were followed for 1.5 years ± 3 months. All babies had a primary aqueductal stenosis (Table 1).

Table (1): Demographic and clinical data of the study group

Item		**Value**
Sex	Male	15(71.4%)
	Female	6(28.6%)
Age (months)	Mean±SD	14.52±5.20
	Median(range)	14(6-24)
	Age <1year	6(28.4%)
	Age 1-2 years	15(71.4%)

The success rate was 66.6% (n = 14) Figure (1) shows the success rate related to age group with a higher success group in the age 1 – 2 years while the higher failure rate was in children below one year. The difference was statistically significant (P=0.010). Complications occurred in 7 babies (33.3%).

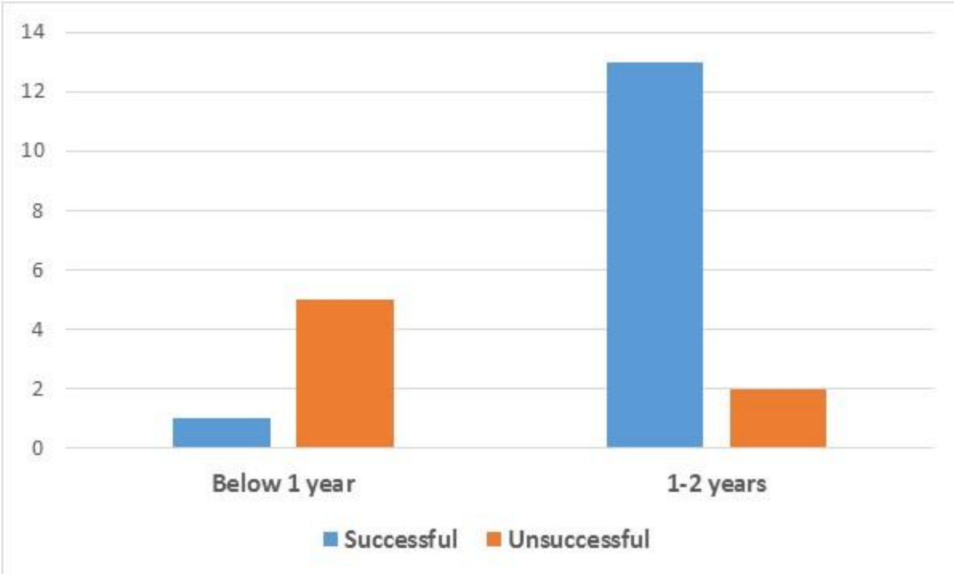

Figure (1): success rate related to the age group

The failure was detected clinically (increase in the head circumference) and radiologically (increase in the ventricular width or periventricular edema) and defined as the need to repeat the ETV or inserting a shunt. In four of six premature babies, ETV failed and needed VP shunt insertion. The mean head circumference was 44 ± 5 cm which matches with more than 2 SD of the median reference for this age.

Five babies (23.8%) needed VP shunt insertion due to ETV failure, 4 males, and one female. One case (4.8%) had a postoperative CSF leak which was controlled by tight resuturing and one case (4.8%) who was the youngest baby, (6 months old) died due to intraventricular hemorrhage, an external ventricular drain was tried but without improvement and the child died in the third day postoperatively. Four weeks after the procedure, 14 children showed a significant clinical improvement with reduction of irritability and the head circumference showed no increase while a postoperative CT to the same children after one month clarified a reduction in the ventricular width and absence of periventricular edema.

Table (2): Individual data of children.

Cases	Age	Gender	Complications
1	18 months	Male	None
2	20 months	Male	None
3	14 months	Female	None
4	7 months	Male	VP shunt inserted
5	24 months	Female	None
6	22 months	Male	None
7	8 months	Male	VP shunt inserted
8	20 months	Female	None
9	6 months	Male	Died
10	18 months	Male	None
11	14 months	Male	VP shunt inserted
12	19 months	Male	None
13	18 months	Female	None
14	13 months	Male	None
15	14 months	Male	None
16	9 months	Female	VP shunt inserted
17	14 months	Male	None
18	9 months	Male	None
19	15 months	Male	CSF leak
20	8 months	Male	VP shunt inserted
21	15 months	Female	None

Case1. 1.5 year old male baby, his mother came complaining of progressive head enlargement, his head circumference was 49 cm (> 97th percentile) his anterior fontanel was 3 cm wide and tense. Figure 2 A shows preoperative CT brain with dilated lateral and third ventricles. Figure2 B one month after the ETV shows prominence of subarachnoid space and reduction in the ventricular size. Figure 2C, one year after the procedure shows the appearance of the basal cisterns and more reduction in the ventricular width.

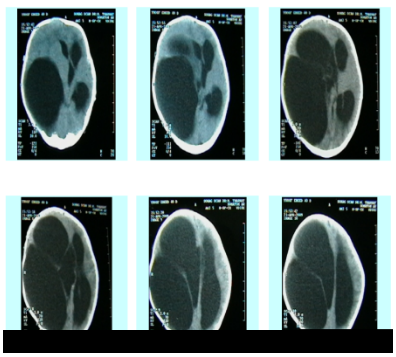

Figure 2A: Preoperative CT

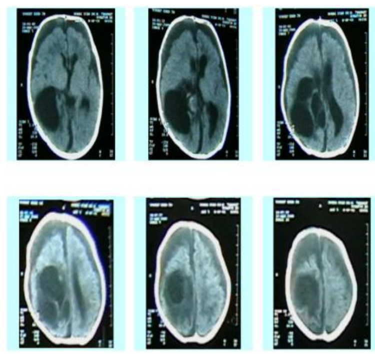

Figure 2B: one month postoperatively

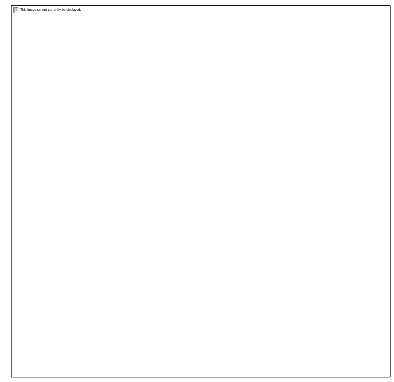

Figure 2C: One year postoperatively

Case 2: Female baby, 15 months old, her mother was complaining of enlarged head size. Her head circumference was 47 cm (larger than 95% percentile), Ant fontanel was 3.5 fingers. Figure 3A shows preoperative CT brain with dilated lateral ventricles with ballooning of the third ventricle. One month after the ETV (figure 3B) CT shows reduction of lateral and third ventricle and follow-up CT brain after one year (Figure 3C) shows more reduction of the ventricular size toward its normal size.

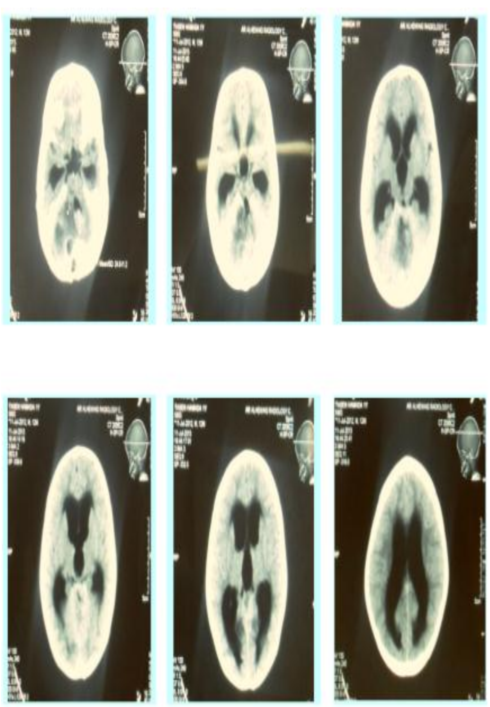

Figure 3 A: Preoperative CT

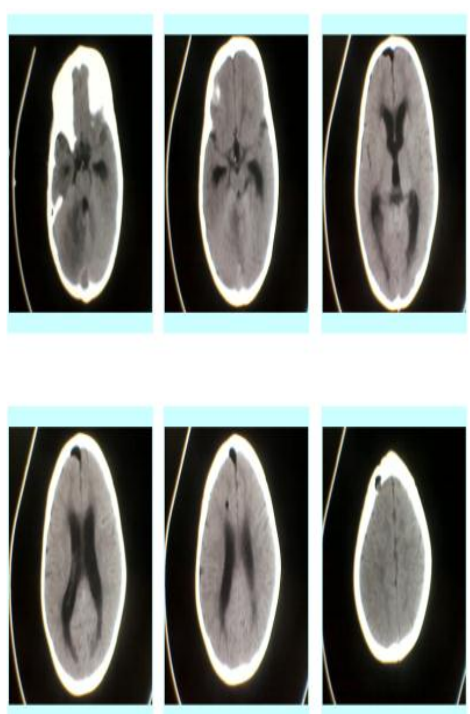

Figure 3 B: after one month postoperatively

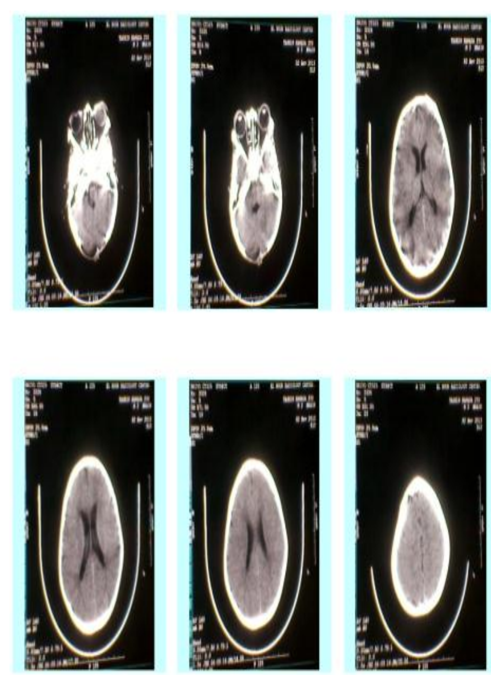

Figure 3 C: after one year postoperatively

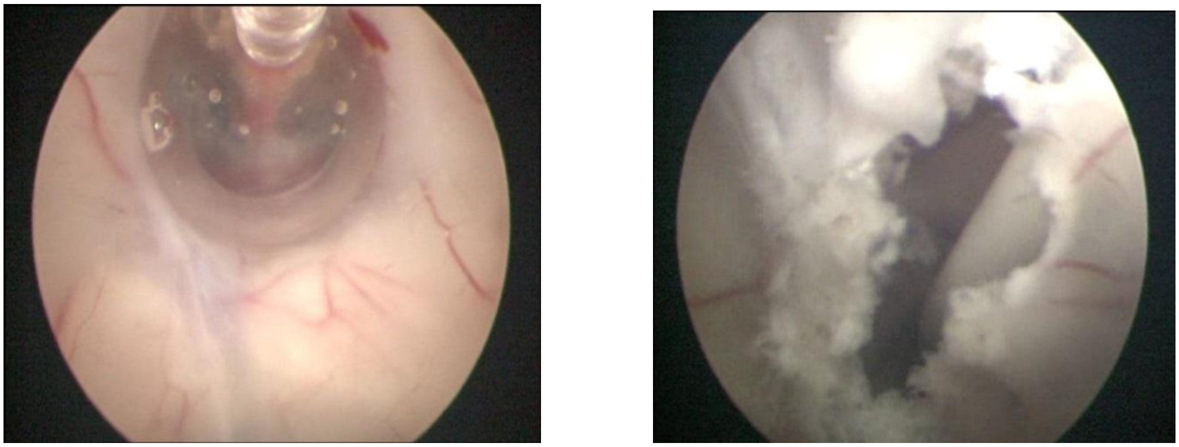

Figure (4): (A) Opening the ventriculostomy and widening using Fogarty catheter and (B) ensuring free CSF flow to the basal cisterns

Discussion:

ETV has become a well-established, effective and safe technique in treating hydrocephalus with different etiologies. However ETV done for children below two years with hydrocephalus is still controversial issue as many authors reported higher failure rates than older children [11, 12]. Our success rate was 66.6% compared to another study [13] with 43%, and L.fani et al. study with 42.5% [14] and 53% in Beems et al. study [15]. All patients in this study had a primary aqueductal stenosis compared to other studies that involved babies with post hemorrhagic, post infection or babies with spina bifida. However, Beems et al. [15] classified children into subgroups according to etiology with a high success rate in hydrocephalus secondary to aqueductal stenosis (87%) if compared to the results of hydrocephalus secondary to myelomeningocele.

Consequently, they assumed that outcome is related more to the etiology rather than the age. Results of this study had the same result with other studies [8, 16]. On the other hand, other studies [14, 17] reported mainly the age factor as the most determinant factor in the outcome with no influence of the etiology, they stated that children below 6 months and preterm babies had open skull sutures and soft skull bones leading to higher compliance and low fontanel pressure which caused ETV failure. In our study, we report that both factors have an influential role on the success of the ETV. This is similar to other studies [9, 18-20]. Our study didn’t include children below 6 months as the failure is very high in this age group. Treating hydrocephalus secondary to aqueductal stenosis has a varying degree of success from 60 % up to 75% according to some authors [21-23].

Post-hemorrhagic and post infection aqueductal stenosis have a higher ETV failure rate according to authors [11]. We had one patient with CSF leak after the ETV (4.8%) compared to another study [14] that had 20% of patients with CSF leak; this may be due to involvement of infants below 6 months with a higher failure rate. The crucial point here is to detect whether the ETV would be successful if applied to children in this age group. Kulkarni and his colleagues [24] tried to answer this fundamental question and put the endoscopic third ventriculostomy score (ETVSS) shown in table (3) to predict the success rate of ETV, with scores ranging from 0 (extremely poor outcome of ETV) to 90 (extremely high chance of ETV success).

We suggest that ETVSS can be used to select good candidates for ETV. ETV technique has a steep learning curve. However, sometimes, ETV becomes a difficult technique in patients with distorted anatomical landmarks especially in babies who have spina bifida [25]. Some anatomical variations were reported in the literatures like shallow prepontine cistern, narrow foramen magnum, thick floor of the third ventricle and small sized third ventricle [4]. Our advice to avoid this is careful examination of the preoperative MRI.

Overall, ETV has more serious morbidity if compared to shunt system, however, these complication are reduced minimally with experienced hands [26].

Hydrocephalic children got benefit more from the ETV as they avoid the complications of shunting for long period [27]. Sufianov and his colleagues assumed that ventriculostomy opening tends to close in the thick ventricular floor [22]. This finding is harmonious with another study [28], which postulated that the tendency to form new membranes is higher in infants than in older patients. On the other hand, some Korean authors suggested that placement of the shunt tube with ETV in the same session carries a higher success rate (> 80%), however, the patient would have the same shunt complications and dependence [17]. In another study, they reported that ETV failure is due to closure of the stoma in about 10% of cases in infants. This closure may be due to less developed subarachnoid space which hinders the CSF absorption with a higher tendency to form new membranes with gliosis [12].

Table (3): ETV scoring system

Score	Age	Etiology	Previous Shunt
0	< 1 month	Post infection	Previous shunt
10	< 1 month	-	No previous shunt
20	1month to < 6 months	Myelomeningocele – non tectal brain tumor - IVH	
30	6 months to < 1 year	Aqueductal stenosis - Tectal brain tumor	
40	1 – 10 years	-	
50	≥ 10 years		

Conclusion:

Our overall success rate was 66.6%. Both age and the cause of hydrocephalus are crucial for the success of ETV. ETV provides the advantages of shunt independence and avoidance of shunt complications. Failure of ETV is higher in premature babies. ETVSS is important in predicting the outcome of ETV and we strongly recommend applying it preoperatively.

The authors have no conflict of interests and no financial support

Acknowledgment:

Authors would like to thank Professor Roshdy Elkhayat for his great efforts and strong support in this humble work.

References:

1. Decq, P., et al., A history of ventricular neuroendoscopy. World Neurosurg, 2013. **79**(2 Suppl): p. S14 e1-6.
2. Guiot, G., Ventriculo-cisternostomy for stenosis of the aqueduct of Sylvius. Puncture of the floor of the third ventricle with a leucotome under television control. Acta Neurochir (Wien), 1973. **28**(4): p. 275-89.
3. Costa Val, J.A., et al., Third ventriculostomy in infants younger than 1 year old. Childs Nerv Syst, 2012. **28**(8): p. 1233-5.
4. Gangemi, M., et al., Endoscopic third ventriculostomy for hydrocephalus. Minim Invasive Neurosurg, 1999. **42**(3): p. 128-32.
5. Di Rocco, C., et al., Endoscopic third ventriculostomy in the treatment of hydrocephalus in pediatric patients. Adv Tech Stand Neurosurg, 2006. **31**: p. 119-219.
6. Garcia, L.G., et al., Endoscopic Third Ventriculostomy Success Score (ETVSS) predicting success in a series of 50 pediatric patients. Are the outcomes of our patients predictable? Childs Nerv Syst, 2012. **28**(8): p. 1157-62.
7. Faggin, R., et al., Hydrocephalus in infants less than six months of age: effectiveness of endoscopic third ventriculostomy. Eur J Pediatr Surg, 2009. **19**(4): p. 216-9.
8. Cinalli, G., et al., Failure of third ventriculostomy in the treatment of aqueductal stenosis in children. Neurosurg Focus, 1999. **6**(4): p. e3.
9. Hayashi, N., et al., Clinical features in patients requiring reoperation after failed endoscopic procedures for hydrocephalus. Minim Invasive Neurosurg, 2000. **43**(4): p. 181-6.
10. Hopf, N.J., et al., Endoscopic third ventriculostomy: outcome analysis of 100 consecutive procedures. Neurosurgery, 1999. **44**(4): p. 795-804; discussion 804-6.
11. Buxton, N., et al., Neuroendoscopic third ventriculostomy in patients less than 1 year old. Pediatr Neurosurg, 1998. **29**(2): p. 73-6.

12. Mohanty, A., et al., Failed endoscopic third ventriculostomy in children: management options. Pediatr Neurosurg, 2002. **37**(6): p. 304-9.
13. Baldauf, J., et al., Endoscopic third ventriculostomy in children younger than 2 years of age. Childs Nerv Syst, 2007. **23**(6): p. 623-6.
14. Fani, L., et al., Endoscopic third ventriculocisternostomy in hydrocephalic children under 2 years of age: appropriate or not? A single-center retrospective cohort study. Childs Nerv Syst, 2013. **29**(3): p. 419-23.
15. Beems, T. and J.A. Grotenhuis, Is the success rate of endoscopic third ventriculostomy age-dependent? An analysis of the results of endoscopic third ventriculostomy in young children. Childs Nerv Syst, 2002. **18**(11): p. 605-8.
16. Elbabaa, S.K., et al., Endoscopic third ventriculostomy for obstructive hydrocephalus in the pediatric population: evaluation of outcome. Eur J Pediatr Surg, 2001. **11 Suppl 1**: p. S52-4.
17. Kim, S.K., K.C. Wang, and B.K. Cho, Surgical outcome of pediatric hydrocephalus treated by endoscopic III ventriculostomy: prognostic factors and interpretation of postoperative neuroimaging. Childs Nerv Syst, 2000. **16**(3): p. 161-8; discussion 169.
18. Balthasar, A.J., et al., Analysis of the success and failure of endoscopic third ventriculostomy in infants less than 1 year of age. Childs Nerv Syst, 2007. **23**(2): p. 151-5.
19. Fritsch, M.J., et al., Endoscopic third ventriculostomy in infants. J Neurosurg, 2005. **103**(1 Suppl): p. 50-3.
20. Gorayeb, R.P., S. Cavalheiro, and S.T. Zymberg, Endoscopic third ventriculostomy in children younger than 1 year of age. J Neurosurg, 2004. **100**(5 Suppl Pediatrics): p. 427-9.
21. Ogiwara, H., et al., Endoscopic third ventriculostomy for obstructive hydrocephalus in children younger than 6 months of age. Childs Nerv Syst, 2010. **26**(3): p. 343-7.
22. Sufianov, A.A., G.Z. Sufianova, and I.A. Iakimov, Endoscopic third ventriculostomy in patients younger than 2 years: outcome analysis of 41 hydrocephalus cases. J Neurosurg Pediatr, 2010. **5**(4): p. 392-401.
23. Yadav, Y.R., et al., Endoscopic third ventriculostomy in infants. Neurol India, 2006. **54**(2): p. 161-3.
24. Kulkarni, A.V., et al., Predicting who will benefit from endoscopic third ventriculostomy compared with shunt insertion in childhood hydrocephalus using the ETV Success Score. J Neurosurg Pediatr, 2010. **6**(4): p. 310-5.
25. Teo, C. and R. Jones, Management of hydrocephalus by endoscopic third ventriculostomy in patients with myelomeningocele. Pediatr Neurosurg, 1996. **25**(2): p. 57-63; discussion 63.
26. Siomin, V., et al., Endoscopic third ventriculostomy in patients with cerebrospinal fluid infection and/or hemorrhage. J Neurosurg, 2002. **97**(3): p. 519-24.
27. Elgamal, E.A., et al., Endoscopic third ventriculostomy for hydrocephalus in children younger than 1 year of age. Childs Nerv Syst, 2011. **27**(1): p. 111-6.
28. Javadpour, M., et al., The impact of endoscopic third ventriculostomy on the management of newly diagnosed hydrocephalus in infants. Pediatr Neurosurg, 2001. **35**(3): p. 131-5.

الملخص العربي

استخدام المنظار الجراحي لحالات استسقاء المخ في الاطفال

مقدمة البحث:

أصبح استخدام المنظار الجراحي لحالات استسقاء المخ بديلا مناسبا لعمليات تركيب صمامات المخ وان كان بعض الاطباء متخوفين من نسبة فشل استخدامه في الاطفال الاقل من عمر سنتين لعدم اكتمال تكون الاغشية المحيطة بالمخ في هذا العمر.

هدف البحث:

تسجيل نسبة نجاح تلك العمليات في الاطفال محل الدراسة للاعمار أقل من عامين.

طريقة البحث:

تم اجراء البحث على عدد واحد وعشرين طفلا كان أغلبهم من الذكور في العمر ما بين ست شهور وسنتين كانوا يعانون من استسقاء أولي بالمخ في الفترة ما بين ابريل ٢٠١٢ الى سبتمبر ٢٠١٤ وتم تسجيل النتائج .

نتيجة البحث:

اربعة عشر طفلا تم اجراء العملية لهم بنجاح بنسبة حوالي ٦٦.٦% بينما خمسة من الاطفال احتاجوا لتكرار استخدام المنظار وطفل قد توفي في اليوم الخامس بعد اجراء المنظار.

ملخص البحث:

يمكن استخدام المنظار الجراحي بأمان في الاطفال من هم دون العامين ولكن بعد الاختيار الدقيق لهذه الحالات لتجنب المضاعفات المتوقعة.