Journal of Clinical Neuroscience xxx (2018) xxx–xxx

Contents lists available at ScienceDirect

Journal of Clinical Neuroscience

journal homepage: www.elsevier.com/locate/jocn

Review article

Endoscopic third ventriculostomy for shunt malfunction in children: A review

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ARTICLE INFO

Article history:
Received 24 July 2017
Accepted 4 February 2018
Available online xxxx

Keywords:
Endoscopic third ventriculostomy
Shunt
Shunt malfunction
Hydrocephalus

ABSTRACT

Endoscopic third ventriculostomy (ETV) is increasingly used in place of shunt revision for shunt malfunction (secondary ETV). This review provides a comprehensive overview of preoperative, operative and postoperative considerations for patients undergoing a secondary ETV. Preoperatively, patient selection is vital and there is evidence that secondary ETV is more effective than primary ETV in certain hydrocephalic aetiologies. Operative considerations include use of neuronavigation and consideration of surgeon technical experience due to anatomical differences that are likely to accompany chronic shunting, management of existing shunt hardware and the use of temporary external ventricular drains or short/long-term ventricular access devices. Postoperatively, there are varying institutional practices with regards to ICP monitoring and length of follow-up after discharge. Finally, this review examines the slit ventricle syndrome as a special case requiring a different approach.

1. Introduction

Endoscopic third ventriculostomy (ETV) is commonly used to manage patients with obstructive hydrocephalus, either as a first-line procedure (primary ETV), or in the setting of shunt malfunction (secondary ETV). Although ventriculostomy for shunt malfunction has been described since the 1960s, surgeons were largely reluctant to use the technique until the advent of refined neuroendoscopic techniques in the late 1990s, when its use became more widespread [1–4].

There are several important factors that surgeons performing a secondary ETV should consider. These can be broadly divided based on clinical course into preoperative, operative and postoperative factors. Preoperatively, the choice of CSF diversion procedure is most important. Scoring systems have been developed to aid clinician decision making, particularly with regards to the likelihood of ETV success [5]. Aetiology of hydrocephalus is a particularly important prognostic indicator. Operative factors include: surgeon technical experience - distorted ventricular anatomy can arise from chronic shunting; management of existing shunt hardware - whether to leave it in, ligate it or remove it entirely; and whether to anticipate ETV failure, most likely in the early postoperative period, by employing an external ventricular drain or ventricular access device. Postoperative factors include the decision to use ICP monitoring and follow-up after hospital discharge.

In this study, we review the literature to provide a comprehensive understanding of secondary ETV.

2. Methods

MEDLINE (PubMed interface) was queried using combinations of the following terms: endoscopic third ventriculostomy, ventriculostomy, ETV, shunt malfunction, shunt failure. Articles were limited to the English language. The search was performed until December 2016. For analysis of secondary ETV outcome, articles meeting the following criteria were included: (1) presentation of clinical outcomes with secondary ETV; (2) inclusion of only paediatric (<18) patients or a predominantly paediatric age group; (3) adequate sample size (n > 10).

3. Preoperative factors

3.1. Efficacy

Studies reporting outcomes with secondary ETV in children are shown in Table 1. Outcomes were available for 519 patients included in 15 observational studies, with a mean age of 9.8 years

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https://doi.org/10.1016/j.jocn.2018.02.012
0967-5868/

Table 1
Outcomes after secondary ETV [4,6–19]. Results from our institution were presented in the study by Stovell *et al.* (highlighted in grey). NR = not reported; AS = aqueductal stenosis; IVH = intraventricular haemorrhage; C-SB: chiari/spina-bifida; mo = months.

Study	Sample size, n						Secondary cause		Mean Age, years (range)	Mean follow-up, months (range)	Shunt-free, n (%)	Complications
	Total	AS	Tumour	Meningitis	IVH	C-SB	Mal-function	Infection				
Jones 1990 Sydney, Australia 1979-1988	14	7	2	1	0	3	NR	NR	12 (9mo-17)	42 (2-120)	8 (57.1%)	0 (0.0%)
Jones 1996 Sydney, Australia 1979-1988	14	0	0	0	0	14	12	2	NR	NR	13 (92.9%)	NR
Teo 1996 Arkansas, USA 1978-1995	55	0	0	0	0	55	NR	NR	11 (1w - 32)	32 (NR)	46 (83.6%)	NR
Cinalli 1998 Paris, France 1974-1995	30	10	7	5	4	0	17	13	8.7 (NR)	104 (6-186)	23 (76.7%)	4 (13.3%)
Beems 2002 Nijmegen, Netherlands Up to 2001	13	2	NR	NR	4	2	NR	NR	5 (3d-18)	50 (6-119)	8 (61.5%)	0 (0.0%)
Siomin 2002 Multi-centre 1993-2000	29	0	0	16	13	0	NR	NR	6.9 (0-65)	22 (7-120)	24 (82.8%)	NR
Buxton 2003 Nottingham, UK 1994-2001	88	27	4	6	12	13	88	0	14 (2mo-76)	36 (1-72)	46 (52.3%)	14 (15.9%)
O'Brien 2005 Multi-centre 1998-2005	63	19	6	4	7	20	49	14	20 (9mo-69)	49 (7-64)	44 (70.0%)	5 (7.9%)
Bilginer 2009 Ankara, Turkey 2002-2007	45	21	6	9	4	3	38	7	12.2 (1-30)	30 (12-60)	36 (80.0%)	0 (0.0%)
Marton 2010 Treviso, Italy 1995-2008	22	4	3	3	7	0	20	2	6.7 (4mo-14)	64 (12-122)	14 (63.6%)	0 (0.0%)
Neils 2013 Illinois, USA 2004-2009	20	6	1	0	3	7	20	0	11.5 (7mo-29)	NR	14 (70.0%)	0 (0.0%)
Brichtova 2013 Brno, Czech Republic 2001-2011	42	15	0	7	15	5	NR	NR	9.5 (NR)	NR	29 (69.0%)	2 (4.8%)
Tamburrini 2013 Rome, Italy 2001-2007	14	0	0	0	0	14	14	0	2.7 (NR)	79 (65-94)	9 (64.0%)	NR
Zhao 2016 Shanghai, China 2005-2014	37	14	9	8	3	0	27	10	1.8 (8mo-3)	3 (NR)	22 (59.5%)	0 (0.0%)
Stovell 2016 Liverpool, UK 1998-2006	**33**	**10**	**1**	**2**	**10**	**10**	**25**	**8**	**6.9 (7d-15.5)**	**53 (1-190)**	**18 (55%)**	**0 (0.0%)**

(95% CI 7.9–11.8 years). The overall pooled efficacy of secondary ETV was 68.2% over a mean follow-up period of 37 months (range 1–190 months) [4,6–19]. The reported efficacy varies between studies due to differences in patient baseline characteristics. ETV is effective in patients presenting with shunt malfunction, as demonstrated by its success in patients with up to 22 prior shunt revisions [4,6,11].

3.2. Aetiology of hydrocephalus

Table 2 shows the impact of aetiology of hydrocephalus on the efficacy of primary and secondary ETV. For aqueductal stenosis and tumours, there was no significant difference in success rate between primary and secondary ETV (OR = 1.19, 95% CI 0.46–3.11; p = 0.74). Secondary ETV had a significantly higher success rate than primary ETV in patients with hydrocephalus due to haemorrhage or infection (OR = 5.79, 95% CI 2.46–13.61; p < 0.001) and chiari malformation (OR = 5.57, 95% CI 2.81–11.00; p < 0.001). These findings are perhaps expected as long-term CSF diversion can induce a state of acquired aqueductal stenosis, for which the efficacy of ETV is similar to the obstructive setting.

3.3. Complication rate

The pooled complication rate of secondary ETV was low at 6.1% across all studies. Reported complications were homologous to primary ETV, including haemorrhage, infection, transient cranial nerve palsies and closure of the stoma site. The complication rates of primary and secondary ETV are similar [12,20], though there are isolated reports of a higher complication rate with the latter [21].

Please cite this article in press as: Waqar M et al. Endoscopic third ventriculostomy for shunt malfunction in children: A review. J Clin Neurosci (2018), https://doi.org/10.1016/j.jocn.2018.02.012