

# HCRN Update: “No shunt for you!” Really?



**Curtis J. Rozzelle, MD**

*Department of Neurosurgery, Pediatric Section;  
University of Alabama at Birmingham School of  
Medicine*



# Disclosures

- *My section chief paid for the trip so there were no personal “Conflict’s of Interest.”*
- *Hydrocephalus Clinical Research Network operations are funded by the Hydrocephalus Association*



# Introduction

- HCRN overview
- Benjamin Warf, MD
- ETV+CPC
- HCRN ETV+CPC Research Plan
- Retrospective Review
- ETV+CPC Training
- CHoA Early Experience
- Next Steps



- Multi-site collaborative clinical research effort
  - Initially 5 busy pediatric neurosurgery programs
  - Now grown to 9 participating centers
- Collect, analyze, & report on high volume rigorous studies to improve diagnosis, treatment, and outcomes for HCP patients
- Provide research-based evidence to support a standard of care.
- [www.hcrn.org](http://www.hcrn.org)

# HCRN Projects

- Shunt Registry
- Shunt Infection QI Protocol
- SOPPH
- Ultrasound Study
- VINOH
- Entry Site Study
- ETV+CPC Study

# Benjamin Warf, MD\*



\*2012 MacArthur Fellow ([www.macfound.org](http://www.macfound.org))

# CURE Children's Hospital: Uganda



# Comparison of endoscopic third ventriculostomy alone and combined with choroid plexus cauterization in infants younger than 1 year of age: a prospective study in 550 African children

**BENJAMIN C. WARE, M.D.**

*CURE Children's Hospital of Uganda, Mbale, Republic of Uganda*

*Object.* The aim of this prospective study was to determine whether, and in which patients, the outcome for bilateral choroid plexus cauterization (CPC) in combination with endoscopic third ventriculostomy (ETV) was superior to ETV alone.

*Methods.* A total of 710 children underwent ventriculoscopy as candidates for ETV as the primary treatment for hydrocephalus. The ETV was accomplished in 550 children: 266 underwent a combined ETV–CPC procedure and 284 underwent ETV alone. The mean and median ages were 14 and 5 months, respectively, and 443 patients (81%) were younger than 1 year of age. The hydrocephalus was postinfectious (PIH) in 320 patients (58%), nonpostinfectious (NPIH) in 152 (28%), posthemorrhagic in five (1%), and associated with myelomeningocele in 73 (13%). The mean follow up was 19 months for ETV and 9.2 months for ETV–CPC. Overall, the success rate of ETV–CPC (66%) was superior to that of ETV alone (47%) among infants younger than 1 year of age ( $p < 0.0001$ ). The ETV–CPC combined procedure was superior in patients with a myelomeningocele (76% compared with 35% success,  $p = 0.0045$ ) and those with NPIH (70% compared with 38% success,  $p = 0.0025$ ). Although the difference was not significant for PIH (62% compared with 52% success,  $p = 0.1607$ ), a benefit was not ruled out (power = 0.3). For patients at least 1 year of age, there was no difference between the two procedures (80% success for each,  $p = 1.0000$ ). The overall surgical mortality rate was 1.3%, and the infection rate was less than 1%.

*Conclusions.* The ETV–CPC was more successful than ETV alone in infants younger than 1 year of age. In developing countries in which a dependence on shunts is dangerous, ETV–CPC may be the best option for treating hydrocephalus in infants, particularly for those with NPIH and myelomeningocele.

**KEY WORDS • hydrocephalus • myelomeningocele • endoscopic third ventriculostomy • choroid plexus cauterization • developing country • pediatric neurosurgery**



# Long-term outcome for endoscopic third ventriculostomy alone or in combination with choroid plexus cauterization for congenital aqueductal stenosis in African infants

## Clinical article

BENJAMIN C. WARF, M.D.,<sup>1,2</sup> SARAH TRACY, B.A.,<sup>3</sup> AND JOHN MUGAMBA, M.D.<sup>4</sup>

*<sup>1</sup>Department of Neurosurgery, Children's Hospital Boston; <sup>2</sup>Department of Global Health and Social Medicine, Harvard Medical School, Boston; <sup>3</sup>University of Massachusetts School of Medicine, Worcester, Massachusetts; and <sup>4</sup>CURE Children's Hospital of Uganda, Mbale, Uganda*

**Object.** The authors have previously reported on the overall improved efficacy of endoscopic third ventriculostomy (ETV) combined with choroid plexus cauterization (CPC) for infants younger than 1 year of age. In the present study they specifically examined the long-term efficacy of ETV with or without CPC in 35 infants with congenital aqueduct stenosis treated at CURE Children's Hospital of Uganda during the years 2001–2006.

**Methods.** Infants with congenital aqueductal stenosis were treated during 2 distinct treatment epochs: all underwent ETV alone, and subsequently all underwent ETV-CPC. Prospectively collected data in the clinical database were reviewed for all infants with an age < 1 year who had been treated for hydrocephalus due to congenital aqueductal stenosis. Study exclusion criteria included: 1) a history or findings on imaging or at the time of ventriculostomy that suggested a possible infectious cause of the hydrocephalus, including scarred choroid plexus; 2) an open aqueduct or an aqueduct obstructed by a membrane or cyst rather than by stenosis; 3) severe malformations of the cerebral hemispheres including hydranencephaly, significant segments of undeveloped brain, or schizencephaly; 4) myelomeningocele, encephalocele, Dandy-Walker complex, or tumor; or 5) previous shunt insertion. The time to treatment failure was analyzed using the Kaplan-Meier method to construct survival curves. Log-rank (Mantel-Cox) and Gehan-Breslow-Wilcoxon tests were used to determine whether differences between the 2 treatment groups were significant.

**Results.** Thirty-five patients met the study criteria. Endoscopic third ventriculostomy alone was performed in 12 patients (mean age 4.7 months), and combined ETV-CPC was performed in 23 patients (mean age 3.5 months). For patients without treatment failure, the mean and median follow-ups were, respectively, 51.6 and 48.0 months in the ETV group and 31.2 and 26.4 months in the ETV-CPC group. Treatment was successful in 48.6% of the patients who underwent ETV alone, as accurately predicted by the Endoscopic Third Ventriculostomy Success Score (ETVSS), and in 81.9% of the patients who underwent ETV-CPC ( $p = 0.0119$ , log-rank test;  $p = 0.0041$ , Gehan-Breslow-Wilcoxon test; HR 6.42 [95% CI 1.51–27.36]).

**Conclusions.** Combined ETV-CPC is significantly superior to ETV alone for infants younger than 1 year of age with congenital aqueductal stenosis. The fact that the outcome for ETV alone was accurately predicted by the ETVSS suggests that these results are applicable in developed countries.

(<http://thejns.org/doi/abs/10.3171/2012.4.PEDS1253>)

# HCRN ETV+CPC Research Plan

- Retrospective Review
- Investigator Training (hands-on)
- Prospective Cohort Study
- Randomized Controlled Trial
  - ETV+CPC vs. VPS for initial mgmt of infant HCP
  - Multiple outcome measures
    - Surgical
    - Neuropsychological/Developmental
    - Cost-effectiveness

# ETV+CPC Retrospective Review\*

- Pop.: All ETV+CPC, 7 sites, 2000-12; n=43
  - Patients <2 yrs; n=36, prior shunt in 13)
    - Premie IVH – 9
    - Aqueductal Stenosis – 8
    - Myelomeningocele – 4
    - Other - 15
- Primary Outcome
  - Time to failure (success rate)
  - Compared with 758 contemporaneous VPS recipients

[\\*J Neurosurg Pediatr.](#) 2014 Jul 4:1-6

# ETV+CPC Retrospective Review\*

- Results:
  - 52% successful @ 2 yrs
  - Slightly worse actuarial survival than VPS ( $p=0.012$ )
  - No major complications
    - 5% infection rate
    - 3 Deaths (1 due to seizure; 2 non-neurologic)
  - Success trends with degree of CPC completed ( $>90\%$ :  $n=11$ ,  $p=0.0501$ )
    - $>90\%$  DoS in 2012 (10/20 vs. 1/16;  $p=0.009$ )
    - Flex-scope  $\rightarrow$   $>90\%$  ( $p<0.001$ )

[\\*J Neurosurg Pediatr.](#) 2014 Jul 4:1-6

# HCRN ETV+CPC Investigator Training

- Needs
  - Hands-on instruction/practice
  - Repetition
- Limitations
  - Time
  - Liability
  - Instructors





# HCRN ETV+CPC Investigator Training



# CURE Children's Hospital: Uganda





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# HCRN ETV+CPC Investigator Training

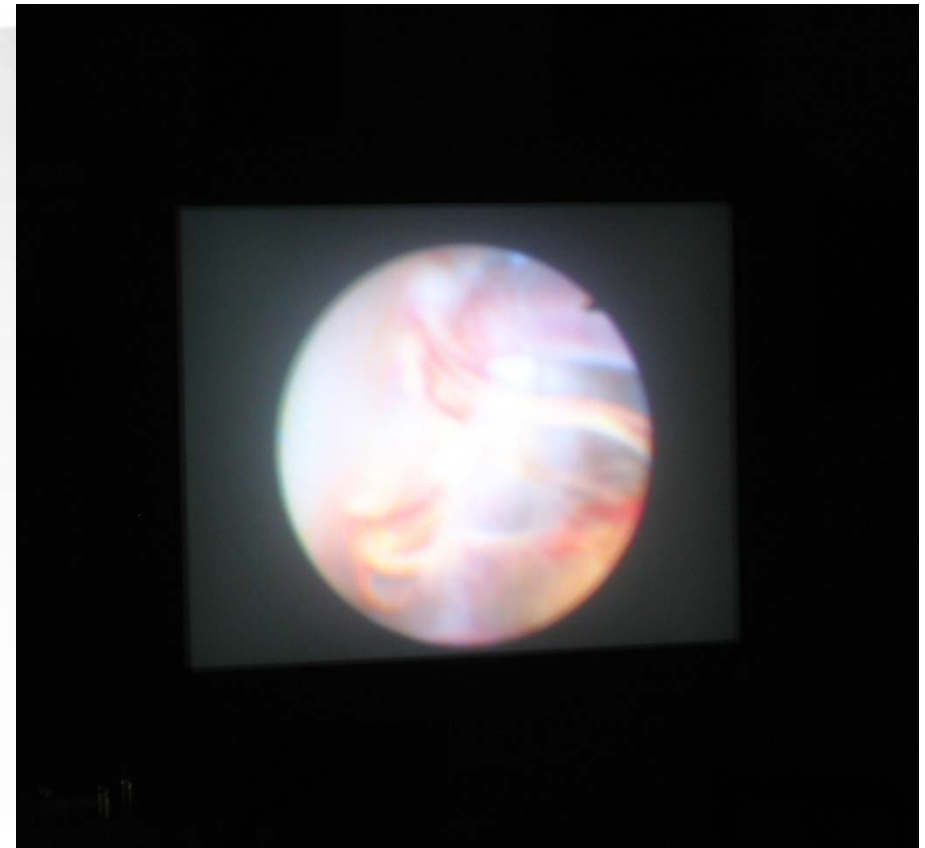


# HCRN ETV+CPC Investigator Training

- ETV+CPC
- Endoscopic Third Ventriculostomy
- Choroid Plexus Coagulation
  - Foramen of Munro to temporal horn
  - Both sides (septostomy prn)
- “Shunt-free” rate 60-80% per etiology of HCP
- Chabra shunt for failures



# HCRN ETV+CPC Investigator Training



# CHoA ETV+CPC Early Experience

- Consider/offer ETV/CPC for
  - All new onset HCP <24 mos. of age
  - “Difficult” shunt patients with ventriculomegaly
- Equipment and technique “Warf style”
- Clinical and imaging surveillance same as VPS



# VIDEO INTERLUDE

# CHoA ETV+CPC Early Experience

- Feb., 2013 – present
- 74 procedures in patients <2y.o.
  - 75 primary treatment
    - Premie-IVH = 29
    - MMC = 15
    - Aqueductal Stenosis = 10
    - Other = 21 (Cyst, Post-infectious, Congenital Communicating, Post-trauma)
- No mortality or major neurologic morbidity.
- Major complication - Diabetes Insipidus x 2



# CHoA ETV+CPC Early Experience

Etiology	# Attempted	# Shunted	Success Rate
Preemie-IVH	29	19	34%
Myelomeningocele	15	4	73%
Aqueductal Stenosis	10	2	80%
Other	21	8	62%
<b>Primary Total</b>	<b>75</b>	<b>33</b>	<b>56%</b>
Secondary	3	2	33%

# Next Steps

- Build on early ETV+CPC experience
- Train more HCRN neurosurgeons
- Follow through!
  - Prospective cohort
  - RCT
- Avoid “rush to judgment”

