Effective Health Care

Pediatric Hydrocephalus

Results of Topic Selection Process & Next Steps

The nominator, Congress of Neurological Surgeons is interested in a new evidence review on pediatric hydrocephalus to inform an update of their clinical practice guideline.

Because limited original research addresses the nomination, a new review is not feasible at this time. No further activity on this nomination will be undertaken by the Effective Health Care (EHC) Program.

Topic Brief

Topic Name: Pediatric hydrocephalus, #749

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Conflict of Interest: None of the investigators have any affiliations or financial involvement that conflicts with the material presented in this report.

Introduction

Hydrocephalus is a condition in which the amount of cerebrospinal fluid (CSF) in the brain is excessive. Pediatric hydrocephalus may be caused by genetic defects, or may be acquired after birth due to head injury, tumors, meningitis, hemorrhage, or premature birth complications that result in CSF flow being blocked from exiting the ventricles or being blocked when in the cisterns. The enlarged ventricles can lead to “potentially harmful pressure on the tissues of the brain.”(NINDS, 2018)

Although the prevalence estimates are low (1-2 babies per 1,000 born (NINDS, 2018)), the burden is high. First, the mortality rate, even after surgical treatment, is high, particularly for pediatric patients in the initial years after surgery (Gmeiner et al., 2017). Additionally, the health and financial burdens are significant. A 2008 study noted that “children with hydrocephalus have a chronic illness and use a disproportionate share of hospital days and healthcare dollars in the US.” (Simon, 2008) Upon analyzing the pediatric hospital discharges in 1997, 2000, and 2003 in the Healthcare Cost and Utilization Project Kids' Inpatient Databases (KID), they determined that, per year, pediatric hydrocephalus resulted in “38,200-39,900 admissions, 391,000-433,000 hospital days, and total hospital charges of $1.4-2.0 billion.”

Using a shunt to move the CSF flow to another cavity is the most common treatment for hydrocephalus (NINDS, 2018). Some of the risks of this treatment include infection, over- or underdrainage of CSF, valve failure, or shunt blockage. In addition, as the child grows, and their brain gets larger, the pressure within the brain increases. This change in pressure can require the shunt valve to be adjusted to a different resistance level. This may potentially be accomplished without shunt replacement if the shunt is equipped with a programmable, adjustable valve; however, if the valve is fixed, then the shunt must be surgically replaced.

One of the most commonly used shunts are the Ventriculoperitoneal Shunting (VPS) shunts. A significant potential complication of overdrainage due to VPS is Slit Ventricle Syndrome (SLS). Its name comes from the small, slit-like appearance of the ventricles upon imaging. SLS is most severe in children, as their brains are increasing in size over time, and the size increase can lead to extreme intracranial pressure (Health, 2018). Diagnosis of SLS is difficult, and there is uncertainty about the best treatment option.

Endoscopic Third Ventriculostomy (ETV) is another treatment option for hydrocephalus. This option could avoid the need for shunting, and thus potentially prevent complications such as SVS oston Children's Hospital, 2018). ETV involves making an opening on the third ventricle floor to allow trapped CSF to drain. ETV is sometimes combined with Choroid Plexus Cauterization (CPC), which involves shrinking the CSF producing choroid plexus via electrical current.

Nominator and Stakeholder Engagement:

The nomination was broad and covered all nine parts of the CNS guideline on this topic as well as several new areas. After consultation with the nominator we narrowed this workup to these new areas. These are:

Key Questions:

KQ1: For children ≤ 1year of age with hydrocephaly, is Endoscopic Third Ventriculostomy (ETV) *with* choroid plexus cauterization (CPC) more effective than ETV *without* CPC at treating hydrocephalus and avoiding the requirement for shunt placement?

KQ2: For children ≤ 21 years of age with a ventriculoperitoneal shunt (VPS) and headache symptoms, what are the most reliable criteria to diagnose slit ventricle syndrome (SVS) and differentiate from other headache causes?

KQ3: For children ≤ 21 years of age with a VPS and headache symptoms, what are the comparative effectiveness and harms of using programmable valves compared to usual care (fixed, unadjustable valves) to prevent SVS?

KQ4: For children ≤ 21 years of age with a VPS and headache symptoms, what is the comparative effectiveness and harms of treatments for SVS?

To define the inclusion criteria for the key questions we specify the population, interventions, comparators, and outcomes (PICO) of interest (Table 1).

Table 1. Key Questions and PICOTS

Key Questions	KQ1	KQ2	KQ3	KQ4
Population	• Newborn to 1 year of age with hydrocephalus	• Newborn to 21 years of age with headache symptoms with VPS for hydrocephaly treatment	• Newborn to 21 years of age with VPS for hydrocephaly treatment	• Newborn to 21 years of age with VPS for hydrocephaly treatment and with SVS
Interventions	• ETV with CPC	• Radiographic tests, such as Magnetic Resonance imaging (MRI), Computerized tomography (CT), and Ultrasound • Clinical assessment	• Programmable valves with adjustable valve resistance	• Adjusting programmable valve resistance • Adding new programmable valve • Adding new fixed valve with higher resistance
Comparators	• ETV without CPC	• Other diagnostic tests or criteria	• Fixed, unadjustable valves	• Other treatment
Outcomes	• Avoiding shunt placement	• Correct treatment • Unnecessary surgery • Delayed necessary surgery	• SVS diagnosis • No SVS diagnosis	• Resolution of SVS • Resolution of headaches
Setting/Study Design	• RCT • Cohort, Case Series ≥ 30 patients	• RCT • Cohort, Case Series ≥ 30 patients	• RCT • Cohort, Case Series ≥ 30 patients	• RCT • Cohort, Case Series ≥ 30 patients

Abbreviations: KQ: key question; RCT: randomized controlled trial; PICOS: Population, Intervention, Comparator, Outcome, Study Design; SVS: Slit Ventricle Syndrome; VPS: Ventriculoperitoneal Shunt; ETV: Endoscopic Third Ventriculostomy; CPC: Choroid Plexus Cauterization

Methods

To assess topic nomination for priority for a systematic review or other AHRQ EHC report, we used a modified process based on established criteria. Our assessment is hierarchical in nature, with the findings of our assessment determining the need for further evaluation. Details related to our assessment are provided in Appendix A.

1. Determine the *appropriateness* of the nominated topic for inclusion in the EHC program.
2. Establish the overall *importance* of a potential topic as representing a health or healthcare issue in the United States.

3. Determine the *desirability of new evidence review* by examining whether a new systematic review or other AHRQ product would be duplicative.
4. Assess the *potential impact* a new systematic review or other AHRQ product.
5. Assess whether the *current state of the evidence* allows for a systematic review or other AHRQ product (feasibility).
6. Determine the *potential value* of a new systematic review or other AHRQ product.

Appropriateness and Importance

We qualitatively assessed the nomination for appropriateness and importance.

Desirability of New Review/Duplication

We searched for relevant high-quality, completed or in-process evidence reviews from the last three years. Databases searched included AHRQ Effective Health Care Program website, Pubmed, Cochrane Collaboration, Canadian Agency for Drugs and Technologies in Health and PROSPERO register of systematic reviews. (Appendix B)

Impact of a New Evidence Review

The impact of a new evidence review was assessed by analyzing the current standard of care, the existence of potential knowledge gaps, and practice variation. We considered whether it was hypothetically possible for this review to influence the current state of practice through various dissemination pathways (practice recommendation, clinical guidelines, etc.).

Feasibility of a New Evidence Review

We conducted a literature search in PubMed from January 2013 to January 2018.

We reviewed all identified titles and abstracts for inclusion and classified identified studies by study design, to assess the size and scope of a potential evidence review.

See Appendix C for the PubMed search strategy and links to the ClinicalTrials.gov search.

Compilation of Findings

We constructed a table with the selection criteria and our assessments (Appendix A).

Results

Appropriateness and Importance

This is an appropriate and important topic. It represents a significant disease burden and high associated costs to consumers, to patients, to health care systems, and to payers. There is uncertainty regarding whether to use programmable vs. nonprogrammable shunts, ETV with or without CPC, and how to diagnose or treat SVS.

Desirability of a New Review/Duplication

For KQs 1, 2, and 4, a new evidence review would not be duplicative of an existing product.

KQ1: There is one review on ETV with and without CPC (Zandian et al., 2014); however, this is not a high quality review. We found other reviews that look at ETV with CPC or ETV without CPC, but do not compare the two.

KQs 2-4: There were no high quality reviews, only a handful of literature reviews.

KQ3: We found a Cochrane Protocol for a Systematic Review that, if completed, would answer KQ3.

See Table 2, Duplication column.

Impact of a New Evidence Review

A new systematic review may have a high level of impact. The standard of care is unclear. Due to the absence of a guideline, there is wide practice variation – some use programmable shunts, and some use fixed shunts; some perform EVT with CPC, and some perform EVT without CPC; Diagnosis varies as well.

Feasibility of a New Evidence Review

A review is not feasible due to the sparsity of the literature. We found a total of six studies, four related to KQ 1, and two related to KQ 4:

- KQ1: We found four non-RCT (Biluts & Admasu, 2016; Kulkarni et al., 2017; Marano et al., 2015; B. C. Warf, 2013).
- KQ4, we found two small studies. (Teo et al., 2013; Zhao et al., 2016)

No studies met criteria for KQ2 or KQ3.

See Table 2, Feasibility column.

Table 2. Key Questions and Results for Duplication and Feasibility

Key Question	Duplication (1/1/2013-1/1/2018)	Feasibility (1/1/2013-1/1/2018)
KQ1: (**ETV/CPC vs. ETV** alone for treating hydrocephalus and avoiding shunting in children less than a year)	Total number of identified systematic reviews: 1 • ETV with and without CPC: 1: (Zandian et al., 2014) (Although there is a SR that covers this KQ, it is not a high quality review.)	Size/scope of review Relevant Studies Identified: 4 Case Series/Cohort ≥ 30 patients: 4: (Biluts & Admasu, 2016; Kulkarni et al., 2017; Marano et al., 2015; B. C. Warf, 2013) Clinicaltrials.gov: 0
KQ2: (**Diagnostic criteria** to diagnose slit ventricle syndrome [SLS])	Total number of identified systematic reviews: 0	Size/scope of review Relevant Studies Identified: 0 Clinicaltrials.gov: 0

Key Question	Duplication (1/1/2013-1/1/2018)	Feasibility (1/1/2013-1/1/2018)
KQ3: (**Programmable vs. fixed valves** to **prevent** SLS)	Total number of identified systematic reviews: 0 • In-process: 2: (Santiago Adalberto Portillo Medina, 2017)(Foster, 2017) (Uncertain that scope will overlap)	Size/scope of review Relevant Studies Identified: 0 Clinicaltrials.gov: 0
KQ4: (**Treatments** for SLS)	Total number of identified systematic reviews: 0	Size/scope of review Relevant Studies Identified: 2 • Case Series/Cohort ≥ 30 patients: 2: (Teo et al., 2013; Zhao et al., 2016) Clinicaltrials.gov: 0

Abbreviations: AHRQ: Agency For Healthcare Research And Quality; KQ: Key Question; RCT: Randomized Controlled Trial; SLS: Slit Ventricle Syndrome; VPS: Ventriculoperitoneal Shunt; ETV: Endoscopic Third Ventriculostomy; CPC: Choroid Plexus Cauterization

Summary of Findings

- Appropriateness and importance: The topic is both appropriate and important.
- Duplication: A new review would not be duplicative of an existing product for KQ1, 2, and 4. No systematic reviews or protocols were identified for these KQs. KQ3, however, would be duplicative based on a protocol in Prospero.
- Impact: A new systematic review would have high impact, given the need for practice guidelines. There is wide practice variation in shunt type and combination of procedures to treat hydrocephalus. A recently published survey of the American Society of Pediatric Neurosurgeons (Kraemer, Sandoval-Garcia, Bragg, & Iskandar, 2017) noted that "there is wide variability in the understanding and management of shunt-dependent hydrocephalus and its complications. Such discrepancies appear to be derived partly from inconsistent familiarity with existing literature but especially from a paucity of high-quality publications." In addition, consensus is lacking on how best to diagnose or treat SLS.
- Feasibility: A review is not feasible due to the sparsity of the literature.

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Appendix A. Selection Criteria Summary

Selection Criteria	Assessment
1. Appropriateness	
1a. Does the nomination represent a health care drug, intervention, device, technology, or health care system/setting available (or soon to be available) in the U.S.?	Yes
1b. Is the nomination a request for a systematic review?	Yes
1c. Is the focus on effectiveness or comparative effectiveness?	Yes
1d. Is the nomination focus supported by a logic model or biologic plausibility? Is it consistent or coherent with what is known about the topic?	Yes
2. Importance	
2a. Represents a significant disease burden; large proportion of the population	Significant disease burden
2b. Is of high public interest; affects health care decision making, outcomes, or costs for a large proportion of the US population or for a vulnerable population	Yes, affects health care decision making, outcomes, or costs for a vulnerable population
2c. Represents important uncertainty for decision makers	Yes, Uncertainty regarding whether to use programmable vs. nonprogrammable shunts, ETV with or without CPC, and how to diagnose or treat SVS.
2d. Incorporates issues around both clinical benefits and potential clinical harms	Yes
2e. Represents high costs due to common use, high unit costs, or high associated costs to consumers, to patients, to health care systems, or to payers	Yes, Represents high associated costs to consumers, to patients, to health care systems, and to payers
3. Desirability of a New Evidence Review/Duplication	
3. Would not be redundant (i.e., the proposed topic is not already covered by available or soon-to-be available high-quality systematic review by AHRQ or others)	Yes, for KQ1, 2, and 4. Although, there are two upcoming (not completed) review that may address KQ 3, none of the other KQs are addressed by existing or forthcoming reviews.
4. Impact of a New Evidence Review	
4a. Is the standard of care unclear (guidelines not available or guidelines inconsistent, indicating an information gap that may be addressed by a new evidence review)?	Yes, The standard of care is unclear, as guidelines have not yet been developed for the topics of this review.
4b. Is there practice variation (guideline inconsistent with current practice, indicating a potential implementation gap and not best addressed by a new evidence review)?	Yes, Due to the absence of a guideline, there is wide practice variation – some use programmable shunts, and some use fixed shunts; some perform EVT with CPC, and some perform EVT without CPC; Diagnosis varies as well.

Selection Criteria	Assessment
5. Primary Research	
5. Effectively utilizes existing research and knowledge by considering: - Adequacy (type and volume) of research for conducting a systematic review - Newly available evidence (particularly for updates or new technologies)	A limited review would be possible. Four studies compared ETV with and without CPC (Biluts & Admasu, 2016; Kulkarni et al., 2017; Marano et al., 2015; B. C. Warf, 2013). No studies met criteria for KQ2 or KQ3. For KQ4, only 2 small studies met criteria (Teo et al., 2013; Zhao et al., 2016).
6. Value	
6a. The proposed topic exists within a clinical, consumer, or policy-making context that is amenable to evidence-based change	Yes. The proposed topic exists within a clinical, consumer, or policy-making context that is amenable to evidence-based change.
6b. Identified partner who will use the systematic review to influence practice (such as a guideline or recommendation)	Yes, Congress of Neurological Surgeons (CNS) will use the SR to create new guidelines.

Abbreviations: AHRQ: Agency for Healthcare Research and Quality; CNS: Congress of Neurological Surgeons (CNS); CPC: Choroid Plexus Cauterization; ETV: Endoscopic Third Ventriculostomy; KQ: Key Question; SR: Systematic Review; SVS: Slit Ventricle Syndrome; VPS: Ventriculoperitoneal Shunt

Appendix B. Search for Evidence Reviews (Duplication)

Listed are the sources searched.

AHRQ: Evidence reports and technology assessments, USPSTF recommendations

Cochrane Systematic Reviews and Protocols
http://www.cochranelibrary.com/

PubMed
PubMed Health http://www.ncbi.nlm.nih.gov/pubmedhealth/

HTA (CRD database): Health Technology Assessments
http://www.crd.york.ac.uk/crdweb/

PROSPERO Database (international prospective register of systematic reviews and protocols)
http://www.crd.york.ac.uk/prospero/

CADTH (Canadian Agency for Drugs and Technologies in Health)
https://www.cadth.ca/

DoPHER (Database of promoting health effectiveness reviews)
http://eppi.ioe.ac.uk/webdatabases4/Intro.aspx?ID=9

ECRI institute
https://www.ecri.org/Pages/default.aspx

Appendix C. Search Strategy & Results (Feasibility)

Ovid MEDLINE(R) Epub Ahead of Print, In-Process & Other Non-Indexed Citations, Ovid MEDLINE(R) Daily and Ovid MEDLINE(R) 1946 to Present

Date Searched: January 8, 2018

	Searches	Results
1	(Third Ventricle/ or third-ventricle.tw,kf.) and (Ventriculostomy/ or (ventriculostom* or ETV).tw,kf.) and (hydrocephalus/su or (hydrocephal* or ventriculomegal* or "aqueductal stenosis").tw,kf.)	980
2	(Choroid Plexus/ or choroid plexus.tw,kf.) and (cautery/ or (cauter* or coagulat* or CPC).tw,kf.)	364
3	and/1-2	63
4	limit 3 to english language	61
5	limit 4 to "review articles"	10
6	limit 5 to yr="2007-2018"	10
7	limit 4 to (clinical trial, phase iii or clinical trial, phase iv or clinical trial or controlled clinical trial or randomized controlled trial)	3
8	limit 7 to yr="2007-2018"	2
9	Slit Ventricle Syndrome/ or "slit ventricle syndrome".tw,kf.	190
10	limit 9 to english language	178
11	limit 10 to "review articles"	28
12	limit 11 to yr="2007-2018"	15
13	limit 10 to (clinical trial, phase iii or clinical trial, phase iv or controlled clinical trial or randomized controlled trial)	0

ClinicalTrials.Gov

Date Searched: January 18, 2018

1) Choroid Plexus: 0 Studies
2) Slit Ventricle Syndrome, Child, Adult: 0 Studies
3) Hydrocephalus, Child, Adult: https://clinicaltrials.gov/ct2/results?cond=Hydrocephalus&term=&cntry=&state=&city=&dist=&Search=Search&flds=aby&recrs=a&recrs=b&recrs=d&recrs=e&recrs=f&age=0&age=1