J Neurosurg Pediatrics (Suppl) 14:35–43, 2014
©AANS, 2014

Pediatric hydrocephalus: systematic literature review and evidence-based guidelines. Part 5: Effect of valve type on cerebrospinal fluid shunt efficacy

Lissa C. Baird, M.D.,[1] Catherine A. Mazzola, M.D.,[2] Kurtis I. Auguste, M.D.,[3] Paul Klimo Jr., M.D., M.P.H.,[4–6] and Ann Marie Flannery, M.D.[7]

[1]Department of Neurological Surgery, Oregon Health & Science University, Portland, Oregon; [2]Division of Pediatric Neurological Surgery, Goryeb Children's Hospital, Morristown, New Jersey; [3]Department of Neurosurgery, University of California, San Francisco, California; [4]Semmes-Murphey Neurologic & Spine Institute; [5]Department of Neurosurgery, University of Tennessee Health Science Center; and [6]Le Bonheur Children's Hospital, Memphis, Tennessee; and [7]Department of Neurological Surgery, Saint Louis University, St. Louis, Missouri

Object. The objective of this systematic review was to examine the existing literature to compare differing shunt components used to treat hydrocephalus in children, find whether there is a superior shunt design for the treatment of pediatric hydrocephalus, and make evidence-based recommendations for the selection of shunt implants when placing shunts.

Methods. Both the US National Library of Medicine PubMed/MEDLINE database and the Cochrane Database of Systematic Reviews were queried using MeSH headings and key words chosen to identify publications comparing the use of shunt implant components. Abstracts of these publications were reviewed, after which studies meeting the inclusion criteria were selected. An evidentiary table was compiled summarizing the selected articles and quality of evidence. These data were then analyzed by the Pediatric Hydrocephalus Systematic Review and Evidence-Based Guidelines Task Force to consider evidence-based treatment recommendations.

Results. Two hundred sixty-nine articles were identified using the search parameters, and 43 articles were recalled for full-text review. Of these, 22 papers met the study criteria for a comparison of shunt components and were included in the evidentiary table. The included studies consisted of 1 Class I study, 11 Class II studies, and 10 Class III studies. The remaining 21 articles were excluded.

Conclusions. An analysis of the evidence did not demonstrate a clear advantage for any specific shunt component, mechanism, or valve design over another.

Recommendation: There is insufficient evidence to demonstrate an advantage for one shunt hardware design over another in the treatment of pediatric hydrocephalus. Current designs described in the evidentiary tables are all treatment options. Strength of Recommendation: Level I, high degree of clinical certainty.

Recommendation: There is insufficient evidence to recommend the use of a programmable valve versus a nonprogrammable valve. Programmable and nonprogrammable valves are both options for the treatment of pediatric hydrocephalus. Strength of Recommendation: Level II, moderate degree of clinical certainty.
(http://thejns.org/doi/abs/10.3171/2014.7.PEDS14325)

Key Words • hydrocephalus • cerebrospinal fluid shunt • practice guidelines • programmable valve • antisiphon device

Hydrocephalus is the most common condition treated by pediatric neurosurgeons. Successful management with cerebrospinal fluid shunt systems began after Nulsen and Spitz placed the first implantable shunt in 1949, using a stainless steel ball-valve system.[34] Over the next 2 decades, shunt systems evolved to include distal slit valves, proximal slit valves, and diaphragm valves. The subsequent development of artificial valves and silicone tubing advanced shunt design dramatically. Simple differential pressure valves were initially engineered followed by a second generation of valves that included autoregulating, adjustable, antisiphon, and gravitational components.

The objective of this systematic review is to examine literature in which differing shunt components used to treat hydrocephalus in children are compared to find whether there is a superior shunt design for the treatment of pediatric hydrocephalus and to make evidence-based recommendations regarding the selection of shunt implants when placing shunts. Currently, many shunt system components are available to the pediatric neurosurgeon, and they function with a variety of pressure, flow, and

Abbreviations used in this paper: AANS = American Association of Neurological Surgeons; CNS = Congress of Neurological Surgeons.

siphon control characteristics. Shunt system design has evolved along with attempts to minimize failure rates. The initial use of simple differential pressure valves led to concerns about the disadvantages of siphoning and associated shunt obstruction, subdural hematoma, slit ventricle syndrome, overdrainage, and craniosynostosis. In an attempt to minimize these complications, antisiphon devices have been developed and integrated into shunt systems as intrinsic to the valve mechanism or as separate devices. The antisiphon device is designed to provide progressive resistance to flow to counteract the siphoning that occurs when negative pressure is exerted with vertical positioning. The later development of programmable valves allowed for purposeful alterations in valve function to be made without a surgical procedure.

The purpose of this evidence-based review is to critically evaluate available data on the efficacy of comparable shunt components to determine if one shunt component is superior to another. Additionally, we created evidence-based recommendations on the selection of shunt components based on the strength of the available data. Most of the available evidence focuses on the comparison of shunt valve designs. Study outcome variables accepted for the purposes of this review included shunt survival, shunt complications, development of slit ventricle syndrome, and development of signs or symptoms of overdrainage.

Methods

Search Criteria

The US National Library of Medicine (PubMed/MEDLINE) and the Cochrane Database of Systematic Reviews were queried for the period January 1966 through March 2012 using MeSH headings and key words relevant to shunt system components as detailed below.

Search Terms

PubMed/MEDLINE

1. ("Cerebrospinal Fluid Shunts"[MeSH]) "Hydrocephalus"[MeSH:noexp]
2. 1 AND (programmable OR nonprogrammable OR non-programmable OR siphon OR antisiphon* OR antisiphon* OR ("differential pressure" OR "fixed pressure") OR valve*)
3. Limit 2 to Child (0–18 years)
4. Limit to English and Humans

Cochrane Database

1. MeSH descriptor Child
2. MeSH descriptor Infant
3. 1 or 2 and (MeSH descriptor Cerebrospinal Fluid Shunts)
4. 3 and (MeSH descriptor Hydrocephalus)
5. (programmable OR nonprogrammable)
6. 4 and 5

Search Results

The search yielded 269 abstracts, which were then reviewed for relevance to the demonstration of superiority of 1 shunt component over another. Forty-three articles were recalled for full-text review. Predetermined inclusion and exclusion criteria were used to review each of these articles in detail. Twenty-two articles were included in the final evidentiary table. Reasons for exclusion of full-text articles included the absence of a valid comparison group (n = 14),[1–3,7–10,12,13,20,25,26,28,35] the absence of a valid outcome variable (n = 4),[14,18,22,32] invalid study design (n = 2),[30,31] and redundant patient population (n = 1) (Fig. 1).[5]

For each article included in the evidentiary table, the study type, summary findings, and major conclusions were recorded, and a preliminary data class was assigned. The Pediatric Hydrocephalus Systematic Review and Evidence-Based Guidelines Task Force met to discuss the ranking of the evidence and the classification of data. Recommendations were then made based on the strength of the data in the evidentiary table (Table 1). In these discussions, if disagreement was encountered among Task Force members, a blinded vote was held and a consensus or majority opinion was reached.

Results

The review process identified 1 Class I study, 11 Class II studies, and 10 Class III studies.

Only one included article was rated as a Class I study, Kestle et al. (2000),[19] in which the investigators performed a randomized controlled trial comparing 3 kinds of valves: all types of standard differential pressure valves, a Delta valve (Medtronic) with an antisiphon mechanism, and an Orbis-Sigma valve (Cordis) with a variable-resistance and flow-limiting mechanism. Three hundred forty-four patients were randomly assigned to a valve type and followed up until the time of first shunt failure. Assessed outcome variables included shunt obstruction, overdrainage, ventricular loculations, and infection. The investigators did not find a significant difference in shunt survival between the 3 valve types in either the short-term (Drake 1998[5]) or extended[19] follow-up.

Eleven Class II studies[4,11,15,21,23,24,27,33,36,41,44] in which differing valve types were compared also failed to demonstrate a superior valve when shunt survival was assessed. Jain et al.[15] (2000) conducted a prospective cohort study in which they compared shunts using a standard differential pressure valve with a Delta (Medtronic) flow-regulating valve. The authors found no significant difference in overall shunt survival (p = 0.72), with a 5-year survival rate of 58.6% for the differential pressure valves and 58.7% for the Delta valves. The authors did note a relative difference between the 2 groups in the incidence of overdrainage and infection. The differential pressure valve was associated with 4 cases of post-shunt subdural effusion or slit ventricle syndrome, while the Delta valve was associated with only 1 case of subdural effusion. The Delta valve group had 3 infections, whereas the differential pressure valve group had no infections. Warf et al.[44] (2005) conducted a prospective randomized trial in which they compared the Codman-Hakim microprecision valve with the more affordable Chhabra valve. Ninety children were evaluated after randomization for shunt malfunction, shunt migration, and wound complication. No significant differences in outcome variables were demon-

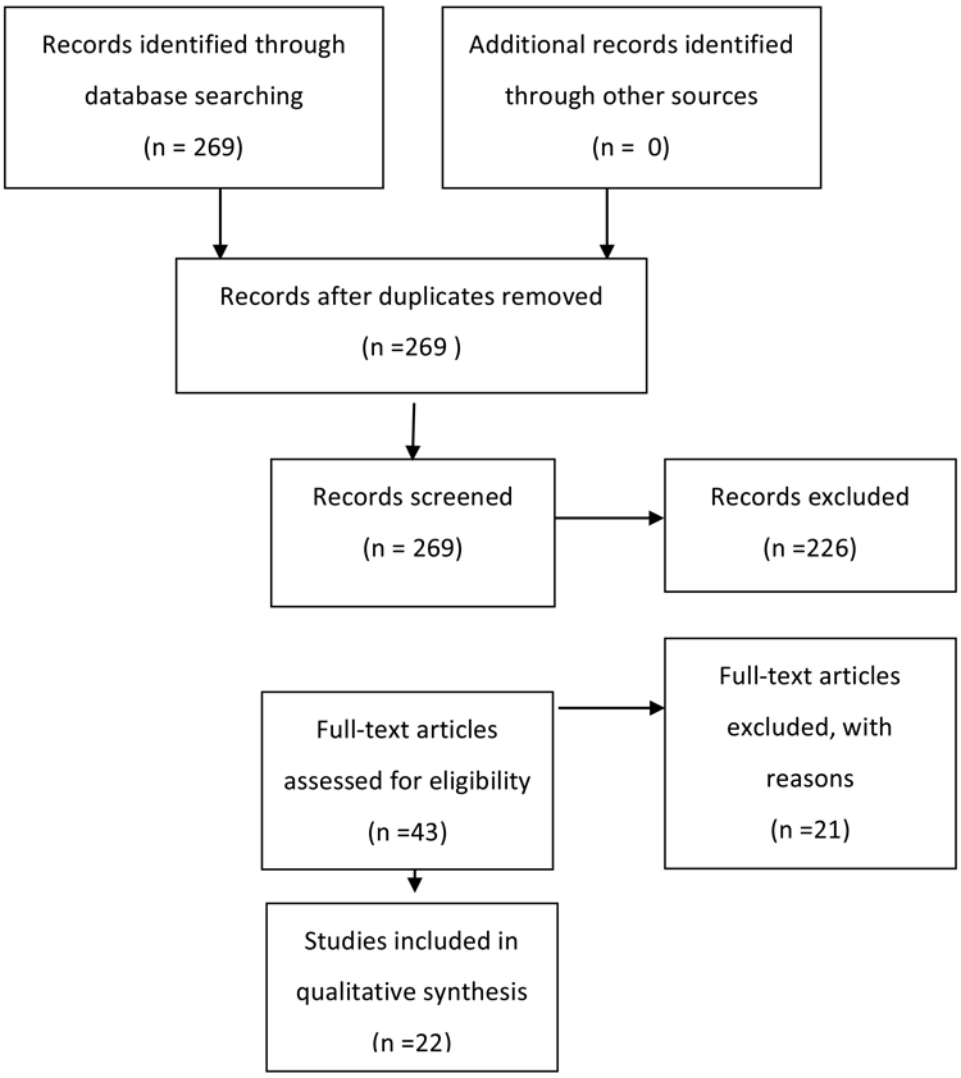

Fig. 1. Flowchart showing the process involved in identifying relevant literature.

strated between the 2 groups. Smely and Van Velthoven (1997) conducted a retrospective cohort review in which they compared 66 infants who underwent placement of a ventriculoperitoneal Cordis Orbis-Sigma valve system with 53 patients who underwent placement of a ventriculoatrial Codman Holter Valve shunt system.[41] Forty-eight percent of patients with the Orbis-Sigma valve required one or more revisions while 98.1% of patients with the Holter Valve required 1 or more revisions ($p < 0.001$). The difference in distal placement of the shunt system is a confounding factor when comparing valve types in this study.

Antisiphon Mechanism

Three Class II studies evaluating the antisiphon mechanism were included in our review. Liniger et al.[23] (2003) studied 27 infants in a prospective cohort study in which a PS Medical medium pressure, flow-controlled valve was compared with a PS Medical 1.0 Delta valve with an antisiphon mechanism. The authors found a lower incidence of slit ventricle syndrome in the Delta valve group (6.25%) than in the flow-controlled valve group (9%); however this finding did not reach statistical significance ($p > 0.99$). The incidence of shunt revision was also lower in the Delta group (0.12 revisions/patient/year) than in the flow-controlled valve group (0.19 revisions/patient/year), a finding that also failed to reach statistical significance ($p = 0.75$). Khan et al.[21] (2010) studied the role of the antisiphon mechanism in a randomized controlled trial. Forty patients undergoing shunt placement were randomized to receive a differential pressure valve with an antisiphon device (Vygon shunt) or a differential valve without an antisiphon device (Chhabra or Ceredrain shunts). Shunt blockage, shunt infection, overdrainage, loculated ventricles, and occipitofrontal circumference were assessed in the 2 groups. No end point variables demonstrated a statistically significant difference. Overdrainage complications occurred in 10% of the patients in the group without an antisiphon device as opposed to 0% in the group with an antisiphon device ($p = 0.48$). A slightly higher infection and obstruction rate was noted in the antisiphon group. In a retrospective cohort study of

TABLE 1: Valve type: summary of evidence*

Authors & Year	Study Description	Data Class, Quality, & Reasons	Results & Conclusions
Kestle et al., 2000	Multicenter randomized trial comparing differential pressure valve, Delta valve, & an Orbis-Sigma valve. 344 pts w/ time to shunt failure as end point.	Class I Randomized controlled trial.	No clear advantage of 1 valve over another.
Khan et al., 2010	Role of ASD. 40 pts randomly assigned to shunt w/ ASD or differential valve.	Class II Prospective, randomized comparative trial. Small study w/ short follow-up (<6 mos).	No overdrainage in ASD group, 2 pts w/ overdrainage in non-antisiphon group; higher occlusion & infection in ASD group. No end point variables reached statistical significance.
Jain et al., 2000	Prospective data from 50 consecutive 1st-time shunt insertions. Comparison of shunt survival between differential pressure & flow-regulating (Delta) valves.	Class II Prospective cohort study.	No significant difference in shunt survival between 2 groups: 5-yr survival = 58.6% in DPV group & 58.7% in Delta valve group. Higher incidence of overdrainage in DPV group & higher rate of infection in Delta valve group, although neither was statistically significant.
Liniger et al., 2003	27 infants w/ flow-control vs antisiphon valves followed for development of slit ventricles & slit ventricle syndrome.	Class II Prospective cohort study.	No significant difference in development of slit ventricles or slit ventricle syndrome between 2 groups. Slit ventricle syndrome developed in 6.25% of pts in the antisiphon group & 9% of pts in the flow-controlled valve group ($p > 0.99$).
Pollack et al., 1999	377 pts randomized to receive the Codman Hakim programmable shunt vs a conventional valve of surgeon's choice & followed for valve explant & shunt failure.	Class II Multicenter randomized controlled trial. Control group not uniform in valve type.	Comparable efficacy & safety w/ no statistically significant difference in shunt survival.
Davis et al., 2000	Retrospective cohort study of 475 pts who underwent VP shunt placement w/ Delta valve w/ antisiphon function or 2 differential pressure valves w/o antisiphon function (Holter-Hausner & Heyer-Schulte). End points included shunt survival & symptomatic subdural fluid collections.	Class II Single-institution retrospective cohort study.	No significant difference in shunt survival at 2-yr follow-up: 65% in Delta group, 66% in Holter-Hasner group, & 64% in Heyer-Schulte valve group. No significant difference in rate of subdural fluid collection between groups.
Hatlen et al., 2012	Retrospective review of 523 pts who underwent 616 shunt surgeries w/ 2-yr min follow-up. Pts w/ programmable valve placement (Strata & Codman Hakim valves) compared w/ nonprogrammable valves (Heyer-Schulte, Spetzer, Delta, & Medtronic). Valve survival was primary end point.	Class II Retrospective cohort study. Data obtained from prospectively collected shunt database.	5-yr survival for nonprogrammable valves (45.8%) was significantly higher than that for programmable valves (19.8%), $p = 0.0001$.
McGirt et al., 2007	279 pts who underwent shunt placement procedures w/ either a programmable (Strata or Codman Hakim) or a nonprogrammable valve (PS Medical Delta) were retrospectively assessed & their cases analyzed for time to shunt malfunction & type of malfunction.	Class II Retrospective cohort review.	Programmable valves associated w/ reduced risk of both overall shunt revision (35% vs 54%, $p = 0.016$) & proximal obstruction (12% vs 28%, $p = 0.006$).
Notarianni et al., 2009	253 pts who underwent shunting procedures w/ either a programmable (Medtronic Strata or Codman Hakim) or a nonprogrammable (pressure-controlled or not specified) valve were retrospectively assessed & their cases analyzed for time to shunt malfunction.	Class II Retrospective review.	Failure rates ($p = 0.11$) were not significantly different btwn shunts w/ programmable valve (76.1%), shunts w/ nonprogrammable valve (80.0%), & shunts w/ nonspecified valve (65.0%).

(continued)

TABLE 1: Valve type: summary of evidence* ***(continued)***

Authors & Year	Study Description	Data Class, Quality, & Reasons	Results & Conclusions
Warf, 2005	90 pts randomized to receive the Chhabra or Codman Hakim shunt as primary treatment for hydrocephalus & 105 pts treated w/ Chhabra shunt after unsuccessful ETV.	Class II Prospective, randomized study. This study was downgraded from a Class I to a Class II study due to nonblinded outcome assessors.	No difference btwn 2 groups in incidence of shunt malfunction, shunt migration, wound complication, or death.
Mangano et al., 2005	189 children who underwent shunt placement w/ either a programmable valve (Strata or Codman-Medos w/ ASD) or a nonprogrammable valve (PS Medical) were retrospectively assessed & their cases analyzed for time to shunt malfunction & CSF protein levels.	Class II Retrospective cohort review, short follow-up (mean 9 mos).	Programmable valves had higher rate of malfunction (11.1% compared to 0%), but the difference did not reach statistical significance.
Smely & Van Velthoven, 1997	Retrospective review of 66 infants treated w/ Cordis Orbis-Sigma Valve compared to 53 children treated w/ Codman Holter Valve ventriculoatrial system.	Class II Retrospective cohort review.	Codman Holter Valve group demonstrated a greater than double risk of shunt complications in comparison to the ventriculoperitoneal Orbis-Sigma valve system. 48.5% of pts w/ Orbis-Sigma valve required 1 or more revisions; 98.1% of pts w/ Holter Valve required 1 or more revisions ($p < 0.001$).
Gruber et al., 1984	Retrospective review of 41 pts in whom there was primary or secondary placement of an ASD to their shunt system. Comparison of clinical course before & after placement of ASD.	Class III Retrospective case series.	Fewer shunt malfunctions after ASD placement. Complication rate per pt was 4 times less frequent & the annual ventricular catheter obstruction rate per pt improved 12 times. No statistical analysis to determine significance.
Kan et al., 2007	244 children who underwent shunt placement w/ either a differential pressure valve, Delta valve, or Orbis-Sigma valve & had 1-yr follow-up data were assessed for development of slitlike ventricles.	Class III Retrospective review.	23 pts developed slitlike ventricles: 10.8% of pts w/ differential pressure valves, 10.5% of pts w/ Delta valves, & 3.6% of pts w/ Orbis-Sigma valves, $p = 0.007$. Children w/ differential pressure or Delta valves were 1.67 times more likely to develop slitlike ventricles than those w/ Orbis-Sigma valves.
Miranda et al., 2011	Retrospective review of shunt survival in 103 pts treated for preterm-related posthemorrhagic hydrocephalus.	Class III Retrospective review.	42 episodes of obstruction. Fixed medium pressure valves were associated w/ a higher rate of obstruction than low pressure valves; only statistically significant in those pts weighing > 2000 g, $p = 0.040$.
Ramadwar et al., 1997	28 pts underwent retrospective comparison of the efficacy of the Delta valve vs the Heyer-Shulte Multi-Purpose valve.	Class III Retrospective review. Small sample.	69% of pts w/ Delta valve & 53% of pts w/ Multi-Purpose valve required revision. Did not reach statistical significance.
Robinson et al., 2002	158 pts whose cases were retrospectively analyzed for significant factors associated w/ shunt malfunction.	Class III Retrospective case series.	Revision rate per yr was 4 times higher for pts w/ no valve or low pressure valve (72% 5-yr failure rate) than for pts w/ medium or high pressure valve (47% 5-yr failure rate), $p = 0.0005$.
Sainte-Rose et al., 1991–1992	Retrospective review of the mechanical complications leading to shunt malfunction in 1719 pts w/ shunted hydrocephalus. Pts were treated w/ distal slit valves or medium pressure proximal valves.	Class III Retrospective case series.	A higher risk of proximal occlusion is associated w/ flanged ventricular catheters ($p < 0.04$); shunts w/ proximal medium pressure valves are less likely to malfunction than shunts w/ distal slit valves ($p < 0.000002$); open-ended distal catheters associated w/ fewer distal obstructions ($p < 0.0003$).

(continued)

TABLE 1: Valve type: summary of evidence* *(continued)*

Authors & Year	Study Description	Data Class, Quality, & Reasons	Results & Conclusions
Tuli et al., 2000	Data prospectively collected on 839 pts who underwent primary shunt insertion. 1183 episodes of shunt failure occurred. Valve types included flow regulated & differential pressure.	Class III Prospective cohort study, post hoc analysis.	No evidence of an association between shunt malfunction & type of shunt hardware.
Virella et al., 2002	Retrospective review of 101 pts who underwent shunt placement w/ a distal slit valve or a Delta level 1 valve w/ an antisiphon component.	Class III Retrospective case series.	No significant differences were found between the distal slit valve & Delta w/ ASD groups in number of revisions, infections, or overdrainage.
Kaiser et al., 1997	Prospective study comparing conventional medium valve w/ Delta level 1 valve in 25 infants younger than 6 mos.	Class III Prospective randomized. Poor description of study design & data.	No difference in number of revisions. Fewer proximal revisions in Delta Valve group. No determination of significance from described data.
Serlo et al., 1986	Retrospective review of consecutive series of 148 children treated w/ shunt placement procedures w/ either the Pudenz-Heyer valve or the Cordis Hakim valve.	Class III Retrospective review.	No significant differences in efficacy. Tendency toward increased rate of catheter rupture in pts w/ Pudenz-Heyer valve & increased rate of slit ventricles in pts w/ Cordis Hakim valve. The higher patency rate of the Pudenz-Heyer valve was statistically significant ($p < 0.001$).

* ASD = antisiphon device; DPV = differential pressure valve; ETV = endoscopic third ventriculostomy; pt = patient; VP = ventriculoperitoneal.

475 patients, Davis et al.[4] (2000) assessed shunt survival and the development of subdural collection in patients treated with a Delta valve shunt with antisiphon function and in patients treated with one of 2 differential pressure valves without antisiphon control. In a comparison of the 3 groups, no significant difference was found.

The Class III studies that assessed the antisiphon mechanism include a retrospective review by Gruber et al.[6] (1984), in which the authors evaluated 41 patients before and after primary or secondary placement of an antisiphon device. In the secondary placement group fewer complications and proximal catheter obstructions were noted after placement of such a device. However, no statistical analysis was provided by the authors to demonstrate the significance of their findings. In a retrospective cohort review of 101 patients who underwent shunt placement, Virella et al.[43] (2002) reported no significant differences between patients who underwent placement of a distal slit valve and patients who underwent placement of a Delta valve with an antisiphon component. The authors assessed the number of revisions, infections, and evidence of overdrainage, and reported that 31% of patients in the distal slit valve group required a single shunt revision and 8% required a second revision, whereas 30% of patients in the Delta valve group required a single revision and 20% required a second. Kaiser et al.[16] (1997) reported a prospective but incompletely described comparison study between a conventional medium pressure valve and the Delta valve. The authors found no difference in the number of shunt revisions.

Slit Ventricles

Kan et al.[17] (2007) conducted a retrospective review of 244 patients with at least 1 year of follow-up after primary shunt placement with a differential pressure valve, a Delta valve, or an Orbis-Sigma valve. Variables associated with the development of slitlike ventricles included patient age (younger age at insertion was associated with a higher incidence of slitlike ventricles; $p = 0.09$), etiology (trauma, infection, and aqueductal stenosis were associated with a higher incidence of slitlike ventricles), and valve type (10.8% of patients with differential pressure valves, 10.5% with Delta valves, and 3.6% with Orbis-Sigma valves developed slitlike ventricles; $p = 0.007$). This article suggests that a slower reduction in ventricle size and slower flow may lead to larger ventricles after shunt placement. Slit ventricle syndrome was not directly assessed; rather, the radiographic appearance of slitlike ventricles was used as a surrogate outcome.

Programmable Valves

Five Class II studies[11,24,27,33,36] evaluated programmable valves. Pollack et al.[36] (1999) conducted a multicenter randomized controlled trial in which they compared the programmable Codman Hakim valve to the surgeon's choice of any conventional valve. The authors demonstrated comparable efficacy and safety with no statistically significant difference in shunt survival between the experimental and control groups.

Hatlen et al. (2012) published an analysis of program-

mable and nonprogrammable valve survival.[11] The programmable Strata and Codman Hakim valves were compared with multiple nonprogrammable valves and found to have significantly lower survival rates (19.8% vs. 45.8%, p = 0.0001). Another retrospective comparison between programmable valves (Strata or Codman-Medos) and nonprogrammable valves (Medtronic PS Medical) was undertaken by Mangano et al.[24] (2005). In that study 11% of the programmable valves malfunctioned compared with 0% of the nonprogrammable valves. The authors demonstrated a trend toward longer valve survival and shunt survival in the nonprogrammable group; however, neither reached statistical significance. McGirt et al. (2007) retrospectively reviewed 279 patients who had undergone shunt placement procedures involving either a programmable (Strata or Codman Hakim) or nonprogrammable (PS Medical Delta) valve.[27] The authors found that programmable valve placement was associated with a reduced risk of both overall shunt revision (35% vs 54% in the nonprogrammable group; p = 0.016) and proximal shunt obstruction (12% vs 28% in the nonprogrammable group; p = 0.006). Notarianni et al.[33] (2009) found no significant difference in a retrospective review of 253 patients who underwent shunt placement with either a programmable (Strata or Codman Hakim) or nonprogrammable (pressure-controlled or not specified) valve. The failure rate among the programmable valve group was 76.1%, and that among the differential pressure valve group was 80.0% (p = 0.11).

Other Comparison Groups

Several Class III studies comparing variable shunt valves were included in the review. Miranda et al.[29] (2011) describe a retrospective review of 103 patients who received shunts for preterm-related posthemorrhagic hydrocephalus. The authors reported a significantly higher rate of obstruction in patients weighing more than 2000 g who were treated with a fixed medium pressure valve (6 of 8 patients) than in those who were treated with a fixed low pressure valve (12 of 39 patients) (p = 0.040). Contrary findings were reported by Robinson et al. (2002) in a retrospective analysis of shunt malfunction variables in 158 patients.[38] Valve opening pressure was the only significant controllable factor found to be associated with shunt malfunction. The 5-year shunt failure rate was 72% in the no valve or low pressure valve group and 47% in the medium or high pressure valve group (p = 0.0005).

Sainte-Rose et al.[39] (1991) reviewed the charts of 1719 patients with shunted hydrocephalus to assess mechanical complications. These authors found that the flanged ventricular catheter was associated with a higher risk of proximal occlusion (p < 0.04), open-ended distal catheters were associated with fewer distal obstructions (log-rank p < 0.0003), and shunts with proximal medium pressure valves were less likely to malfunction than shunts with distal slit valves (p < 0.000002). Tuli et al.[42] (2000) did not find valve type to be associated with shunt malfunction in a post hoc analysis of a prospective cohort of 839 patients who underwent primary shunt insertions. No association between shunt malfunction and any component of the shunt hardware was reported in that study.

Ramadwar et al.[37] (1997) retrospectively compared the efficacy of the Delta valve with the Heyer-Shulte Multi-Purpose valve in 28 patients. Sixty-nine percent of patients with the Delta valve required revision, compared with 53% of patients with the Multi-Purpose valve. The sample size in that study was small, and the data did not reach statistical significance. In an older paper by Serlo et al.[40] (1986), a retrospective review of 148 children was conducted to compare the Pudenz-Heyer valve with the Cordis Hakim valve. No significant difference was found in overall shunt efficacy, although significance was demonstrated in a higher rate of valve patency on the part of the Pudenz-Heyer valve (p < 0.001).

Excluded Studies

The Task Force excluded 21 articles recalled for full-text review from the final evidentiary table. The majority of excluded papers did not include a comparison group or control group.[1–3,7–10,12,13,20,25,26,28,35] Other reasons for exclusion included invalid study design (questionnaire survey),[30,31] redundant patient population[5] (only the paper with the longest reported follow-up was included), and absence of a valid outcome variable (change in ventricle size, development of spinal canal stenosis, historical description, and frequency of hospital visits).[14,18,22,32]

Conclusions

Recommendation: There is insufficient evidence to demonstrate an advantage of one shunt hardware design over another for the treatment of pediatric hydrocephalus. Current designs described in the evidentiary tables are all treatment options. Strength of Recommendation: Level I, high degree of clinical certainty.

Recommendation: There is insufficient evidence to recommend the use of a programmable valve versus a nonprogrammable valve. Programmable and nonprogrammable valves are both options for the treatment of pediatric hydrocephalus. Strength of Recommendation: Level II, moderate degree of clinical certainty.

The available literature in which one shunt component is compared with another does not demonstrate a clear superiority of one over another. Higher rates of overdrainage were seen with standard differential pressure valves; however, the outcome variables studied in the comparisons of these groups with other shunt mechanisms failed to demonstrate statistical significance. While valves with antisiphon mechanisms may be superior in preventing overdrainage complications, no statistically significant data exist in the current medical literature to support this trend.

The studies assessing programmable versus nonprogrammable valves demonstrated either no statistically significant differences or contrary outcomes, pointing to the need for long-term prospective controlled analysis of this issue. Class III data demonstrating poorer function of distal slit valves in comparison with a proximal valve are described and are consistent with the contemporary decrease in utilization of the former type of shunt system.

Many contemporary valve designs exist despite major deficiencies in long-term clinical evaluation. Well-designed comparison studies with clearly defined outcome

variables and appropriate stratification of patient variables are needed to further investigate the appropriate clinical utilization of these valves. Accessing the necessary patient volume required to reach significance and balancing industry interests with trial integrity may be significant barriers to pursuing needed studies; however, as increasingly expensive and complex valves become available for clinical use, these studies will become imperative.

Acknowledgments

We acknowledge the American Association of Neurological Surgeons (AANS)/Congress of Neurological Surgeons (CNS) Joint Guidelines Committee for the members' reviews, comments, and suggestions; Laura Mitchell, Guidelines Project Manager for the CNS, for her contributions; Pamela Shaw, research librarian, for her assistance with the literature searches; Kevin Boyer for his assistance with data analysis; and Sue Ann Kawecki for her assistance with editing.

Disclosure

The systematic review and evidence-based guidelines were funded exclusively by the CNS and AANS Pediatric Section, which received no funding from outside commercial sources to support the development of this document.

Conflict(s) of Interest: None. All Task Force members declared any potential conflicts of interest prior to beginning work on this evidence review.

Author contributions to the study and manuscript preparation include the following. Conception and design: AANS/CNS Joint Section on Pediatrics. Acquisition of data: all authors. Analysis and interpretation of data: all authors. Drafting the article: Baird. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Flannery. Administrative/technical/material support: all authors. Study supervision: Flannery.

References

1. Ahn ES, Bookland M, Carson BS, Weingart JD, Jallo GI: The Strata programmable valve for shunt-dependent hydrocephalus: the pediatric experience at a single institution. **Childs Nerv Syst 23:**297–303, 2007
2. Arnell K, Eriksson E, Olsen L: The programmable adult Codman Hakim valve is useful even in very small children with hydrocephalus. A 7-year retrospective study with special focus on cost/benefit analysis. **Eur J Pediatr Surg 16:**1–7, 2006
3. Breimer GE, Sival DA, Hoving EW: Low-pressure valves in hydrocephalic children: a retrospective analysis. **Childs Nerv Syst 28:**469–473, 2012
4. Davis SE, Levy ML, McComb JG, Sposto R: The delta valve: how does its clinical performance compare with two other pressure differential valves without antisiphon control? **Pediatr Neurosurg 33:**58–63, 2000
5. Drake JM, Kestle JR, Milner R, Cinalli G, Boop F, Piatt J Jr, et al: Randomized trial of cerebrospinal fluid shunt valve design in pediatric hydrocephalus. **Neurosurgery 43:**294–305, 1998
6. Gruber R, Jenny P, Herzog B: Experiences with the anti-siphon device (ASD) in shunt therapy of pediatric hydrocephalus. **J Neurosurg 61:**156–162, 1984
7. Guthkelch AN: The treatment of infantile hydrocephalus by the Holter valve. **Br J Surg 54:**665–673, 1967
8. Haberl EJ, Messing-Juenger M, Schuhmann M, Eymann R, Cedzich C, Fritsch MJ, et al: Experiences with a gravity-assisted valve in hydrocephalic children. Clinical article. **J Neurosurg Pediatr 4:**289–294, 2009
9. Hahn YS: Use of the distal double-slit valve system in children with hydrocephalus. **Childs Nerv Syst 10:**99–103, 1994
10. Hanlo PW, Cinalli G, Vandertop WP, Faber JA, Bøgeskov L, Børgesen SE, et al: Treatment of hydrocephalus determined by the European Orbis Sigma Valve II survey: a multicenter prospective 5-year shunt survival study in children and adults in whom a flow-regulating shunt was used. **J Neurosurg 99:**52–57, 2003
11. Hatlen TJ, Shurtleff DB, Loeser JD, Ojemann JG, Avellino AM, Ellenbogen RG: Nonprogrammable and programmable cerebrospinal fluid shunt valves: a 5-year study. Clinical article. **J Neurosurg Pediatr 9:**462–467, 2012
12. Hertle DN, Tilgner J, Fruh K, Keinert T, Hagenston AM, Unterberg AW, et al: Reversible occlusion (on-off) valves in shunted tumor patients. **Neurosurg Rev 34:**235–242, 2010
13. Hoekstra A: Artificial shunting of cerebrospinal fluid. **Int J Artif Organs 17:**107–111, 1994
14. Jain H, Natarajan K, Sgouros S: Influence of the shunt type in the difference in reduction of volume between the two lateral ventricles in shunted hydrocephalic children. **Childs Nerv Syst 21:**552–558, 2005
15. Jain H, Sgouros S, Walsh AR, Hockley AD: The treatment of infantile hydrocephalus: "differential-pressure" or "flow-control" valves. A pilot study. **Childs Nerv Syst 16:**242–246, 2000
16. Kaiser GL, Horner E, Marchand S, Jost A: Conventional versus Delta valve in the treatment of hydrocephalus in early infancy. **Eur J Pediatr Surg 7 (Suppl 1):**45–46, 1997
17. Kan P, Walker ML, Drake JM, Kestle JR: Predicting slitlike ventricles in children on the basis of baseline characteristics at the time of shunt insertion. **J Neurosurg 106 (5 Suppl):** 347–349, 2007
18. Keen J: Casey Holter and the Spitz-Holter valve. **Eur J Pediatr Surg 2 (Suppl 1):**5–6, 1992
19. Kestle J, Drake J, Milner R, Sainte-Rose C, Cinalli G, Boop F, et al: Long-term follow-up data from the Shunt Design Trial. **Pediatr Neurosurg 33:**230–236, 2000
20. Kestle JR, Walker ML: A multicenter prospective cohort study of the Strata valve for the management of hydrocephalus in pediatric patients. **J Neurosurg 102 (2 Suppl):**141–145, 2005
21. Khan RA, Narasimhan KL, Tewari MK, Saxena AK: Role of shunts with antisiphon device in treatment of pediatric hydrocephalus. **Clin Neurol Neurosurg 112:**687–690, 2010
22. Kondageski C, Thompson D, Reynolds M, Hayward RD: Experience with the Strata valve in the management of shunt overdrainage. **J Neurosurg 106 (2 Suppl):**95–102, 2007
23. Liniger P, Marchand S, Kaiser GL: Flow control versus antisiphon valves: late results concerning slit ventricles and slit-ventricle syndrome. **Eur J Pediatr Surg 13 (Suppl 1):**S3–S6, 2003
24. Mangano FT, Menendez JA, Habrock T, Narayan P, Leonard JR, Park TS, et al: Early programmable valve malfunctions in pediatric hydrocephalus. **J Neurosurg 103 (6 Suppl):** 501–507, 2005
25. Martínez-Lage JF, Almagro MJ, Del Rincón IS, Pérez-Espejo MA, Piqueras C, Alfaro R, et al: Management of neonatal hydrocephalus: feasibility of use and safety of two programmable (Sophy and Polaris) valves. **Childs Nerv Syst 24:**549–556, 2008
26. Mauer UM, Kunz U: More malfunctioning Medos Hakim programmable valves: cause for concern? Clinical article. **J Neurosurg 115:**1047–1052, 2011
27. McGirt MJ, Buck DW II, Sciubba D, Woodworth GF, Carson B, Weingart J, et al: Adjustable vs set-pressure valves decrease the risk of proximal shunt obstruction in the treatment of pediatric hydrocephalus. **Childs Nerv Syst 23:**289–295, 2007
28. Meling TR, Egge A, Due-Tønnessen B: The gravity-assisted Paedi-Gav valve in the treatment of pediatric hydrocephalus. **Pediatr Neurosurg 41:**8–14, 2005
29. Miranda P, Simal JA, Menor F, Plaza E, Conde R, Botella C: Initial proximal obstruction of ventriculoperitoneal shunt in

patients with preterm-related posthaemorrhagic hydrocephalus. **Pediatr Neurosurg 47:**88–92, 2011
30. Miyake H, Ohta T, Kajimoto Y, Ogawa D: A clinical survey of hydrocephalus and current treatment for hydrocephalus in Japan: analysis by nationwide questionnaire. **Childs Nerv Syst 15:**363–368, 1999
31. Moritake K, Nagai H, Miyazaki T, Nagasako N, Yamasaki M, Sakamoto H, et al: Analysis of a nationwide survey on treatment and outcomes of congenital hydrocephalus in Japan. **Neurol Med Chir (Tokyo) 47:**453–461, 2007
32. Nomura S, Fujii M, Kajiwara K, Ishihara H, Suehiro E, Goto H, et al: Factors influencing spinal canal stenosis in patients with long-term controlled hydrocephalus treated with cerebrospinal fluid shunt. **Childs Nerv Syst 26:**931–935, 2010
33. Notarianni C, Vannemreddy P, Caldito G, Bollam P, Wylen E, Willis B, et al: Congenital hydrocephalus and ventriculoperitoneal shunts: influence of etiology and programmable shunts on revisions. Clinical article. **J Neurosurg Pediatr 4:**547–552, 2009
34. Nulsen FE, Spitz EB: Treatment of hydrocephalus by direct shunt from ventricle to jugular vein. **Surg Forum:**399–403, 1951
35. Piatt JH Jr, Carlson CV: A search for determinants of cerebrospinal fluid shunt survival: retrospective analysis of a 14-year institutional experience. **Pediatr Neurosurg 19:**233–242, 1993
36. Pollack IF, Albright AL, Adelson PD: A randomized, controlled study of a programmable shunt valve versus a conventional valve for patients with hydrocephalus. **Neurosurgery 45:**1399–1411, 1999
37. Ramadwar RH, Carachi R, Young DG: Infantile hydrocephalus: a comparison of the Delta valve and multipurpose valve. **Eur J Pediatr Surg 7 (Suppl 1):**44–45, 1997
38. Robinson S, Kaufman BA, Park TS: Outcome analysis of initial neonatal shunts: does the valve make a difference? **Pediatr Neurosurg 37:**287–294, 2002
39. Sainte-Rose C, Piatt JH, Renier D, Pierre-Kahn A, Hirsch JF, Hoffman HJ, et al: Mechanical complications in shunts. **Pediatr Neurosurg 17:**2–9, 1991–1992
40. Serlo W, von Wendt L, Heikkinen ES, Heikkinen ER: Ball and spring or slit and core valve for hydrocephalus shunting? **Ann Clin Res 18 (Suppl 47):**103–106, 1986
41. Smely C, Van Velthoven V: Comparative study of two customary cerebrospinal fluid shunting systems in early childhood hydrocephalus. **Acta Neurochir (Wien) 139:**875–882, 1997
42. Tuli S, Drake J, Lawless J, Wigg M, Lamberti-Pasculli M: Risk factors for repeated cerebrospinal shunt failures in pediatric patients with hydrocephalus. **J Neurosurg 92:**31–38, 2000
43. Virella AA, Galarza M, Masterman-Smith M, Lemus R, Lazareff JA: Distal slit valve and clinically relevant CSF overdrainage in children with hydrocephalus. **Childs Nerv Syst 18:**15–18, 2002
44. Warf BC: Comparison of 1-year outcomes for the Chhabra and Codman-Hakim Micro Precision shunt systems in Uganda: a prospective study in 195 children. **J Neurosurg 102 (4 Suppl):**358–362, 2005

Manuscript submitted June 25, 2014.
Accepted July 7, 2014.
Please include this information when citing this paper: DOI: 10.3171/2014.7.PEDS14325.

Address correspondence to: Ann Marie Flannery, M.D., Department of Neurological Surgery, Saint Louis University, 3565 Vista Ave., St. Louis, MO 63110. email: flanneam@slu.edu.