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Liquorshunts in der Therapie des kindlichen Hydrozephalus

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Inhaltsverzeichnis

Abkürzungen	4
1) Einleitung	
1.1) Definitionen des Begriffs Hydrozephalus	5
1.2) Pathophysiologie des Hydrozephalus	8
1.3) Ätiologie des kindlichen Hydrozephalus	12
1.4) Symptome und klinische Zeichen	14
1.5) Bildgebende Diagnostik	15
1.6) Neurochirurgische Therapieoptionen	17
1.7) Komplikationen und Outcome	21
1.8) Fragestellungen und Ziele der eigenen Arbeiten	24
2) Eigene Arbeiten	
2.1) Real-time ultrasound guidance for ventricular catheter placement in pediatric cerebrospinal fluid shunts. Beez T, Sarikaya-Seiwert S, Steiger HJ, Hänggi D. Childs Nerv Syst. 2015 Feb;31(2):235-41.	26
2.2) Role of ventriculoperitoneal shunt valve design in the treatment of pediatric hydrocephalus--a single center study of valve performance in the clinical setting. Beez T, Sarikaya-Seiwert S, Bellstädt L, Mühmer M, Steiger HJ. Childs Nerv Syst. 2014 Feb;30(2):293-7.	31
2.3) Prospective review of 30-day morbidity and mortality in a paediatric neurosurgical unit. Campbell E, Beez T, Todd L. Childs Nerv Syst. 2017 Mar;33(3):483-489.	35
2.4) Comparison of quality metrics for pediatric shunt surgery and proposal of the negative shunt revision rate. Beez T, Steiger HJ. World Neurosurg. 2017 Oct. [Epub ahead of print]	40

2.5)	Headache and shunt-related impact on activities of daily life in patients growing up with a ventriculoperitoneal shunt. Beez T, Bellstädt L, Steiger HJ, Sarikaya-Seiwert S. J Neurol Surg A Cent Eur Neurosurg. 2017 Sep. [Epub ahead of print]	46
3)	Diskussion	
3.1)	Technische Aspekte bei der Implantation von Liquorshunts	51
3.2)	Outcome der modernen Therapie des kindlichen Hydrozephalus mit Liquorshunts	53
4)	Zusammenfassung	54
5)	Literaturangaben	55
	Erklärung	75

Abkürzungen

CISS = Constructive interference in the steady state

CT = Computertomografie

ETV = Endoscopic third ventriculostomy, Drittventrikulostomie

FOHR = Frontal occipital horn ratio

HOQ = Hydrocephalus outcome questionnaire

HUI3 = Health utilities index mark 3

MRT = Magnetresonanztomografie

SIKI = Strukturiertes Kopfschmerzinterview für Kinder

SF-36 = 36-Item short form health survey

1) Einleitung

“Perhaps the fundamental lesson from the last 30 years is that hydrocephalus, which appears to be an alluringly simple problem of CSF accumulation, is anything but.”

James M. Drake¹

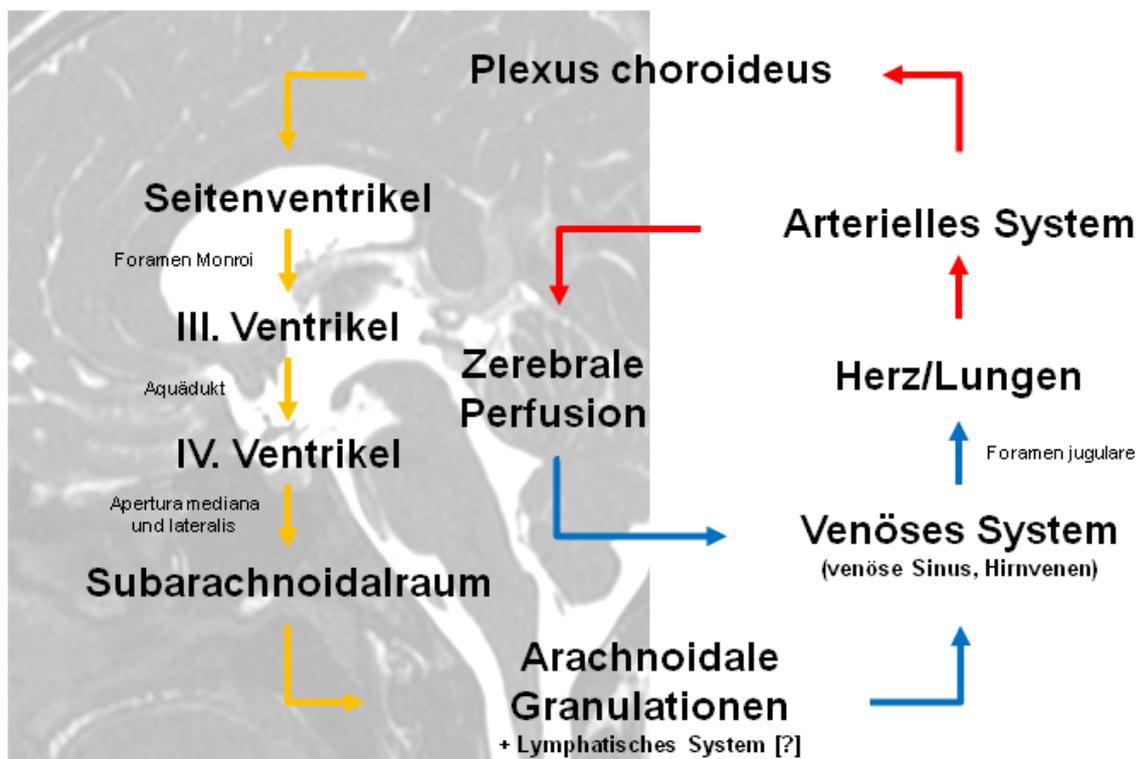
1.1) Definitionen des Begriffs Hydrozephalus

Die Frage nach der Definition des Begriffs Hydrozephalus ist bis in die heutige Zeit Gegenstand wissenschaftlicher Debatten. Angesichts der folgenden Zahlen mag dies verwundern: Im Jahr 2015 wurden in Deutschland fast 7.000 Liquorshunt-Implantationen durchgeführt. In diesem Jahr war Hydrozephalus in 13.000 Fällen die Hauptdiagnose von Patienten, die aus deutschen Krankenhäusern entlassen wurden.² Zum Thema „Hydrozephalus“ finden sich über 30.000 Publikationen in der PubMed-Datenbank der *US National Library of Medicine*. Die Suche nach „Hydrozephalus“ und „Definition“ resultiert hingegen in nur acht Publikationen mit diesem Titel.

Grundlegende Aspekte der modernen Definition des Hydrozephalus wurden bereits Anfang des 20. Jahrhunderts von Dandy formuliert. Er erkannte die große Bedeutung bildgebender Untersuchungen für das Verständnis des Hydrozephalus und führte an seinen Patienten Ventrikulographien nach Injektion von Luft in die Ventrikel des Gehirns durch.³ Er analysierte die zeitabhängige Verteilung der Luft vom Injektionsort in den Ventrikeln bis hin zu den äußeren Liquorräumen und vermutete dort eine Resorption in die systemische Zirkulation. Auf Basis dieser Ergebnisse unterschied Dandy kommunizierende und nicht-kommunizierende Formen des Hydrozephalus

und beschrieb den Verschluss des Aquädukts als ein typisches Beispiel eines nicht-kommunizierenden Hydrozephalus.⁴

Dieses Konzept hat in seinen Grundzügen bis in die heutige Zeit Bestand. Rekate definierte Hydrozephalus als eine aktive Erweiterung des Ventrikelsystems des Gehirns, hervorgerufen durch eine inadäquate Passage des Liquors von seinem Entstehungsort in den Ventrikeln zu seinem Resorptionsort in die systemische Zirkulation.^{5,6} Zudem entwickelte er ein auf technisch-mathematischen Überlegungen basierendes hydraulisches Kreislaufmodell der Liquorzirkulation (Grafik 1), welches in der Folge sowohl *in vitro* als auch an Tiermodellen validiert wurde.⁷ Dieses Modell erlaubt die Erklärung und Untersuchung verschiedener Hydrozephalus-Formen, aber auch Forschung an Erkrankungen dieses Formenkreises im weiteren Sinne, wie zum Beispiel dem *Pseudotumor cerebri*.



Grafik 1. Illustration des hydraulischen Kreislaufmodells, angelehnt an Rekate (Cerebrospinal Fluid Research 2008, 5:2 doi:10.1186/1743-8454-5-2).

Raimondi erkannte die Limitationen, welche eine Definition mit sich bringt, die explizit eine Erweiterung der Ventrikel als obligates Element beinhaltet. Er definierte Hydrozephalus daher als jede pathologische Zunahme des intrakraniellen Liquorvolumens, nicht notwendigerweise innerhalb der Ventrikel, unabhängig vom hydrostatischen Druck.⁸ Dieses Konzept beinhaltet intraparenchymale Formen, wie zum Beispiel das Hirnödem, sowie extraparenchymale Formen. Letztere Kategorie umfasst Erhöhungen des Liquorvolumens sowohl im intraventrikulären Raum als auch in den zisternalen und subarachnoidalen Kompartimenten. Diese recht offene Definition schließt somit auch den externen Hydrozephalus sowie seltene Formen, wie den Niedrigdruckhydrozephalus, mit ein.

Unter Einbeziehung moderner bildgebender Verfahren, insbesondere der Magnetresonanztomografie (MRT), publizierte ein Experten-Panel, die *Hydrocephalus Classification Study Group*, im Jahr 2011 eine Konsensusklassifikation des Hydrozephalus.⁵ Diese Klassifikation basiert weiterhin auf einem Obstruktionsmodell und unterscheidet sechs mögliche Punkte der Obstruktion des Liquorflusses: *Foramina Monroi*, Aquädukt, Ausflussbahn des IV. Ventrikels, spinaler und kortikaler Subarachnoidalraum, arachnoidale Granulationen sowie der Druck des venösen Systems. Dieses Modell lässt eine Korrelation mit den ätiologischen Subtypen des Hydrozephalus sowie den möglichen Therapien zu. Es soll daher als Basis zukünftiger experimenteller und klinischer Studien dienen und eine bessere Vergleichbarkeit von wissenschaftlichen Ergebnissen in diesem Feld ermöglichen.

1.2) Pathophysiologie des Hydrozephalus

Wesentliche Grundzüge des bis heute gültigen Grundverständnisses der pathophysiologischen Vorgänge des Hydrozephalus bzw. des intrakraniellen Druckes im Allgemeinen gehen auf Arbeiten von Monro, Kellie und Burrows im 18. und 19. Jahrhundert zurück. Diese Untersuchungen resultierten in einer von Cushing im frühen 20. Jahrhundert etablierten Doktrin, nach welcher bei geschlossenem Schädel die Summe der Volumina von Gehirn, Blut und Liquor konstant ist, so dass die Volumenzunahme einer dieser Komponenten bzw. das Auftreten einer weiteren Variable zu einer Volumenabnahme der anderen Komponenten führt, da Gehirn und Flüssigkeit nicht komprimierbar sind.⁹ Dieses grundlegende Konzept ignoriert allerdings die Dynamik der einzelnen Komponenten, den zeitlichen Verlauf einer Volumenänderung sowie die Materialeigenschaften insbesondere des Gehirns.

Hakim entwickelte ein Modell, welches die mechanischen Eigenschaften des Materials „Gehirn“ beachtete.¹⁰ Er beschrieb das Gehirn als einen aus offenen Zellen oder Kompartimenten bestehenden „Schwamm“ aus viskoelastischem Material. Über die Zellen stehen der Liquorraum und das venöse System in Verbindung, so dass letztendlich eine Deformierung des theoretisch nicht komprimierbaren Gehirn-Flüssigkeit-Systems möglich wird, welche beim Hydrozephalus beobachtet werden kann.

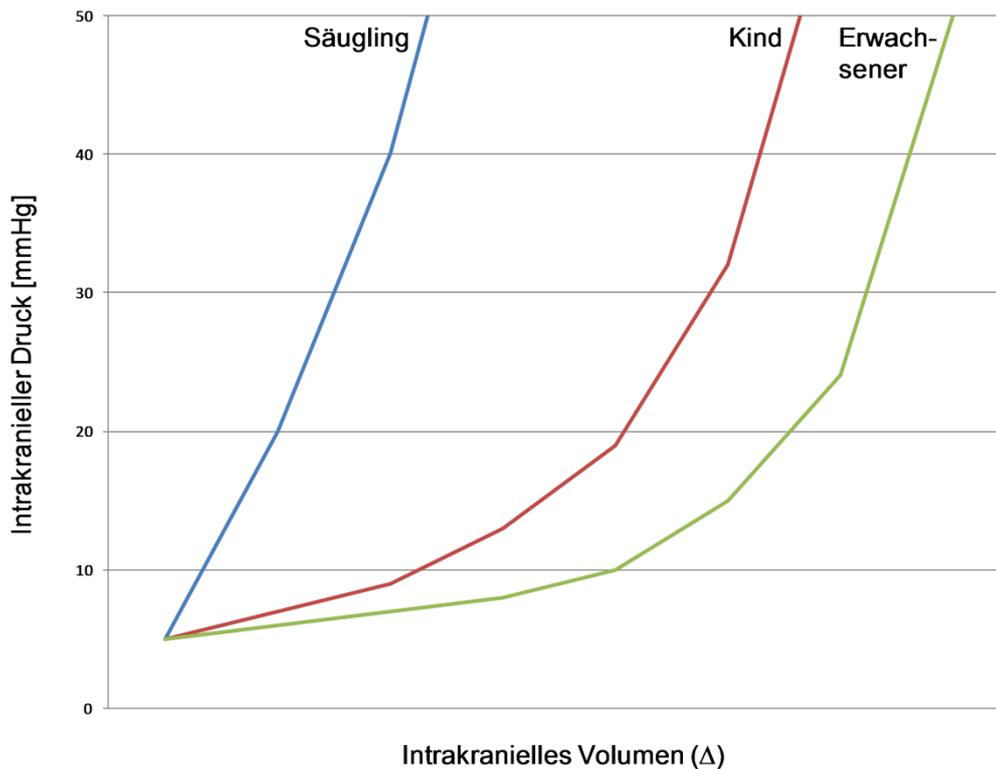
Im weiteren Verlauf fanden die komplexen regionalen Materialeigenschaften des Gehirnparenchyms sowie die komplexe Form der Grenzflächen zwischen Gehirn und Ventrikelsystem zunehmend Beachtung. Nagashima erstellte ein Finite-Elemente-Modell und führte computergestützte Simulationen mit örtlicher und zeitlicher Auflösung durch, welche gut mit klinischen Beobachtungen bei hydrozephalen Patienten, u.a. bei einem siebenjährigen Kind mit akutem Okklusivhydrozephalus,

korrelierten.¹¹ Beispielsweise konnte im Modell gezeigt werden, dass die Parenchymdeformität zunächst das Frontalhorn betrifft und der interstitielle Druckgradient bzw. die Liquordiapese sich zentrifugal vom Ventrikelsystem ausbreitet, das heißt initial vor allem das periventrikuläre Parenchym betrifft. Auch Rekate nutzte mathematische, mechanische und computergestützte Methoden um in ähnlicher Weise das Verhältnis zwischen Ventrikelweite und intrakraniellm Druck bei verschiedenen Formen des Hydrozephalus sowie den Turgor des Gehirns, d.h. die intrinsische Eigenschaft des Gehirns einer Verformung zu widerstehen, zu untersuchen.^{7,12}

Einen wichtigen Beitrag zum Verständnis der *in vivo* Zusammenhänge einzelner Parameter bei hydrozephalen Patienten lieferte Czosnyka durch umfangreiche Daten zu Hirndruck-Monitoring sowie Liquorinfusionsstudien.^{13,14} Er etablierte Basisparameter der Liquordynamik, welche für die Diagnostik eines Hydrozephalus sowie für die Indikationsstellung und Erfolgskontrolle einer Hydrozephalusbehandlung genutzt werden können. Diese Parameter umfassen *resistance to cerebrospinal fluid outflow*, *brain compliance*, *elasticity coefficient*, *estimated sagittal sinus pressure* und *cerebrospinal fluid formation rate*.¹⁵ Beispielsweise konnte durch solche Messungen der *in vivo* Effekt eines Liquorshunts demonstriert werden: Nach Implantation eines Shuntsystems zeigte sich eine Abnahme der *resistance to cerebrospinal fluid outflow* sowie eine Reduktion des mittleren intrakraniellen Drucks.¹⁶ Zudem konnte im Falle einer Shundysfunktion mit Unterdrainage ein erneuter Anstieg dieser Parameter gezeigt werden.¹⁴

Abschließend ist anzumerken, dass sich die meisten der genannten Untersuchungen, Hypothesen und Modelle auf Erwachsene beziehen. Im Gegensatz zu dem bei Erwachsenen geschlossenen Schädel, welcher einen Faktor der Monro-Kellie-Burrows-Doktrin darstellt, ist der Schädel bei Kindern zumindest

bis zum zweiten Lebensjahr aus funktioneller Sicht aufgrund der Fontanellen und Suturen verformbar. Die anteriore Fontanelle ist mit einem mittleren Durchmesser von ca. 2 cm bei Geburt das größte verformbare Areal des kindlichen Schädels. Sie ist bei 96% der Kinder mit einem Alter von 24 Monaten verschlossen.¹⁷ Die kleinere posteriore Fontanelle ist bereits zwischen dem 3. bis 6. Lebensmonat verschlossen. Die im Bereich der Stirn lokalisierte *Sutura metopica* verschließt sich als erste der Schädelnähte mit ca. 12 Monaten, wohingegen die übrigen Nähte teils bis in das Erwachsenenalter zumindest anatomisch nicht komplett verknöchern.¹⁸ Diese anatomischen Verhältnisse führen zu einer erhöhten Compliance des kindlichen Schädeldaches, was bei langsam fortschreitender intrakranieller Raumforderung, z.B. im Sinne eines Hydrozephalus, eine Kompensation durch Zunahme des Kopfumfanges ermöglicht. Bei einer akuten intrakraniellen Raumforderung allerdings reagieren kleine Kinder bereits bei relativ geringeren Volumina mit intrakranielltem Druckanstieg, da ihre Druck-Volumen-Kurve im Vergleich zu älteren Kindern und Erwachsenen steiler verläuft (Grafik 2).



Grafik 2. Theoretische Druck-Volumen-Kurven im altersabhängigen Vergleich. Auffällig ist die deutlich geringere *akute* Volumentoleranz bei jungen Menschen. Es zeigt sich aufgrund des höheren Verhältnisses von Hirnvolumen zu Schädelvolumen ein steilerer ICP-Anstieg bei *akuter* Volumenzunahme. Angelehnt an Shapiro (Shapiro K, Ed., Pediatric Head Trauma, 1983. Futura Publishing, New York, USA) sowie Ropper (N Engl J Med 2012;367:746-52 doi 10.1056/NEJMct1206321).

Wichtige Unterschiede finden sich auch bezogen auf die Physiologie des Liquorsystems bei Kindern: Das Liquorvolumen beträgt bei Neugeborenen ca. 50 ml, zwischen dem 4. und 13. Lebensjahr ca. 90 ml und bei Erwachsenen ca. 150 ml, wovon sich 20% in den Ventrikeln und 80% im kraniellen und spinalen Subarachnoidalraum befinden.¹⁹ Die Liquorproduktion erreicht im ersten Lebensjahr 56% und zweiten Lebensjahr 76% der Produktion eines 15-jährigen Menschen.^{20,21} Während es im ersten Lebensjahr zu einer schnellen altersabhängigen Zunahme der Produktion kommt, flacht der Anstieg jenseits des 1. Lebensjahres ab. Die mittlere Liquorproduktionsrate liegt bei 0,35 ml/min und ist weitgehend unabhängig von temporären Schwankungen des intrakraniellen Druckes.²² Die obere Grenze des normalen intrakraniellen Druckes liegt bei Neugeborenen und Säuglingen bei 5

mmHg, bei Kindern zwischen dem 4. und 16. Lebensjahr bei 13 mmHg und bei Erwachsenen bei 15 mmHg.²³

Möglicherweise bestehen zudem fundamentale Unterschiede in der basalen Physiologie des Liquorsystems zwischen verschiedenen Altersgruppen, die jedoch noch Gegenstand aktueller Forschung sind: Johnston et al. und andere Gruppen konnten tierexperimentell zeigen, dass bei Neonaten die Liquordrainage über extrakranielle lymphatische Wege, insbesondere im Bereich der Hirnnerven sowie der *Lamina cribrosa*, bedeutsamer zu sein scheint als die klassischerweise vermutete Drainage über die arachnoidalen Granulationen in die venösen Sinus.^{24–27}

1.3) Ätiologie des kindlichen Hydrozephalus

Hinsichtlich der Ätiologie des kindlichen Hydrozephalus lassen sich kongenitale von erworbenen Entitäten unterscheiden (Tabelle 1).²⁸ Der kongenitale Hydrozephalus umfasst in seiner strengsten Definition jene Subtypen, welche durch angeborene Fehlbildungen des Gehirns (zum Beispiel Atresie des *Foramen Monroi* oder kongenitale Aquäduktstenose), des kraniozervikalen Übergangs (zum Beispiel Chiari-Malformationen) sowie der Wirbelsäule (zum Beispiel Neuralrohrdefekte) hervorgerufen werden. In gewisser Weise als fließender Übergang zum erworbenen Hydrozephalus werden zudem sekundäre kongenitale Hydrozephalusformen beschrieben. Diese werden ausgelöst durch Ereignisse, welche in der Fetalperiode auftreten und sind damit bereits bei Geburt manifest. Zu diesen auslösenden Faktoren zählen Tumore, Blutungen und Infektionen während der Fetalperiode.

Kongenitaler Hydrozephalus	Erworbener Hydrozephalus
Atresie des Foramen Monroi	Tumor-assoziiert (postnatal)
Kongenitale Aquäduktstenose	Postinfektiös (postnatal)
Neuralrohrdefekte (Hydranzephalie, Enzephalozele, Spina bifida aperta)	Posthämorrhagisch (postnatal)
Dandy-Walker-Malformation	Posttraumatisch (postnatal)
Chiari-Malformationen	
Arachnoidalzysten	
X-chromosomal rezessiver Hydrozephalus	
Sekundärer kongenitaler Hydrozephalus	
Tumor-assoziiert (fetal)	
Postinfektiös (fetal)	
Posthämorrhagisch (fetal)	

Tabelle 1. Einteilung des kindlichen Hydrozephalus nach Ätiologie.

Der erworbene Hydrozephalus hingegen resultiert aus Erkrankungen in der postnatalen Periode. Diese Kategorie umfasst den tumor-assoziierten, posthämorrhagischen, postinfektiösen sowie posttraumatischen Hydrozephalus.

In den westlichen Industrienationen beträgt die Inzidenz des kongenitalen Hydrozephalus 0,5 bis 1 Fälle pro 1.000 Lebendgeburten.²⁹ Der erworbene kindliche Hydrozephalus ist häufiger, mit einer Inzidenz von 3 bis 5 Fälle pro 1.000 Lebendgeburten.^{29,30} In dieser Kategorie ist der posthämorrhagische Hydrozephalus bei Frühgeborenen der häufigste Subtyp. Während für Entwicklungsländer Publikationen zur präzisen Inzidenz fehlen, nimmt man aufgrund von häufiger Mangelernährung mit niedrigem Geburtsgewicht, hoher Infektionsrate und oftmals lückenhafter medizinischer Versorgung sowie fehlender antenataler Diagnostik eine deutlich höhere Inzidenz an.³⁰ Häufigste Ursachen des Hydrozephalus in diesen Ländern sind Neuralrohrdefekte sowie Infektionen.^{30,31}

1.4) Symptome und klinische Zeichen

Die wichtigsten Determinanten der klinischen Symptomatik des kindlichen Hydrozephalus sind die zeitliche Dynamik der Entstehung sowie das Lebensalter des Kindes.

Die Geschwindigkeit der Entstehung eines Hydrozephalus bestimmt darüber, wie effektiv die Kompensationsmechanismen des Gehirns den Liquorüberschuss ausgleichen können. Ein akuter Hydrozephalus resultiert in progredienten Hirndruckzeichen und kann im Extremfall rasch zu Vigilanzminderung bis hin zu Koma durch zerebrale Herniation führen. Im Gegensatz dazu entwickeln sich die Symptome eines chronischen Hydrozephalus oftmals subtil und können sogar in den Status eines *arrested hydrocephalus* übergehen, der einem kompensierten, oft asymptomatischen hydrozephalen Zustand entspricht.³²

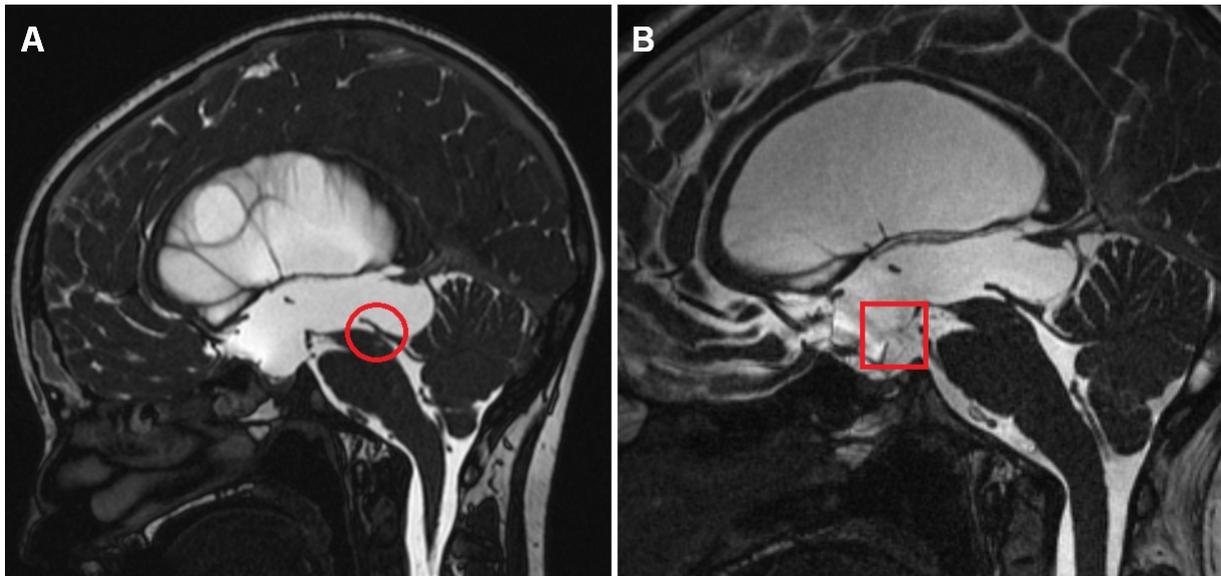
Typische Symptome bei Neugeborenen und Säuglingen sind Irritabilität, Lethargie, Trinkschwäche, Erbrechen und exzessives Schreien insbesondere nachts. Eine vertikale Blickparese kann sich als sogenanntes Sonnenuntergangsphänomen manifestieren. Bei der Inspektion fallen eine gespannte Fontanelle, klaffende Schädelnähte und, insbesondere bei chronischem Hydrozephalus, ein über der Altersnorm liegender bzw. perzentilenflüchtiger Kopfumfang auf.^{33,34} Zudem kann eine prominente Venenzeichnung des Kopfes bestehen. Bei dekompenzierter intrakranieller Druckerhöhung zeigt sich oftmals eine Bradykardie.

Bei älteren Kindern und Jugendlichen gleicht sich das klinische Bild mehr und mehr dem des Erwachsenen an. Es bestehen oftmals morgendlich betonte Kopfschmerzen, Übelkeit und Erbrechen. Doppelbilder aufgrund einer Affektion des *Nervus abducens* sowie Visusminderung aufgrund von Stauungspapillen sind typische okuläre Symptome.

1.5) Bildgebende Diagnostik

In den ersten zwei Lebensjahren kommt dem Ultraschall eine besondere Bedeutung bei der Diagnostik des Hydrozephalus zu, da diese Methode leicht verfügbar ist, keine Strahlenexposition beinhaltet und ohne Sedierung des Kindes erfolgen kann. Andererseits ist die Methode limitiert durch eine starke Untersucherabhängigkeit, eingeschränkte Auflösung sowie begrenzte Beurteilbarkeit insbesondere der hinteren Schädelgrube. Initialdiagnostik sowie Reihenuntersuchungen zur Verlaufskontrolle sind die Haupteinsatzgebiete des kraniellen transfontanellären Ultraschalls in der genannten Altersgruppe. Insbesondere beim posthämorrhagischen Hydrozephalus nach germinaler Matrixblutung des Frühgeborenen sind ultraschalldiagnostische Kriterien etabliert worden, nach welchen über die therapeutische Vorgehensweise entschieden werden kann.³⁵

Goldstandard in der bildgebenden Diagnostik des Hydrozephalus ist die kranielle MRT.³⁶ Die hohe Auflösung sowie die angesichts zahlreicher Spezialsequenzen sehr hohe Aussagekraft und der Verzicht auf ionisierende Strahlung stellen die Stärken der Methode dar, während die aufgrund der langen Untersuchungsdauer und Anfälligkeit für Bewegungsartefakte oftmals notwendige Sedierung des Kindes ein Nachteil ist. In Ergänzung zu konventionellen T1- und T2-gewichteten Sequenzen kann die Detaildarstellung des Liquorsystems mittels *three-dimensional constructive interference in the steady state* (3D CISS) Sequenzen (Grafik 3) deutlich verbessert werden und mittels *cardiac-gated cine phase-contrast imaging* eine nicht-invasive Darstellung des Liquorflusses erfolgen.³⁷



Grafik 3. CISS Sequenzen einer 16 Jahre alten Patientin mit triventrikulärem Hydrozephalus bei Aquäduktstenose. Im präoperativen Bild (A) erkennt man eine kurzstreckige Stenose des Aquädukts (roter Kreis). Im postoperativen Bild (B) nach Drittventrikulostomie sieht man das offene Stoma am Boden des dritten Ventrikels (rotes Viereck). Anzumerken ist, dass der Boden des dritten Ventrikels präoperativ nach unten disloziert war und postoperativ wieder in seine normale Position zurückgekehrt ist.

Die kranielle Computertomografie (CT) ist eine schnelle, leicht verfügbare Methode, welche jedoch mit Strahlenexposition einhergeht und eine begrenzte Auflösung aufweist.¹⁸ Das geschätzte mit einer kraniellen CT eines einjährigen Kindes assoziierte Lebenszeitrisko einer tödlichen strahleninduzierten Krebserkrankung liegt bei 0,07%.³⁸ Einen großen Stellenwert hat die CT dennoch als zeitkritische Notfalluntersuchung. Mittels moderner *low-dose* Protokolle lässt sich die Strahlenexposition um ca. 60% reduzieren, allerdings auf Kosten der Bildqualität.³⁹

Die konventionelle Röntgen-Diagnostik hat bei der Diagnosestellung eines Hydrozephalus keine Bedeutung mehr. Bei Patienten mit Liquorshunts können Röntgen-Aufnahmen eines Verstell-Ventils zum Ablesen der Druckstufe sowie Röntgen-Aufnahmen des Shuntverlaufs bei Verdacht auf Diskonnektion, Dislokation oder Katheterruptur in Einzelfällen sinnvoll sein. Routinemäßige Röntgen-Aufnahmen des Shuntverlaufs werden nicht mehr empfohlen, da nur in weniger als 1% der Fälle eine therapeutische Konsequenz daraus entsteht.⁴⁰

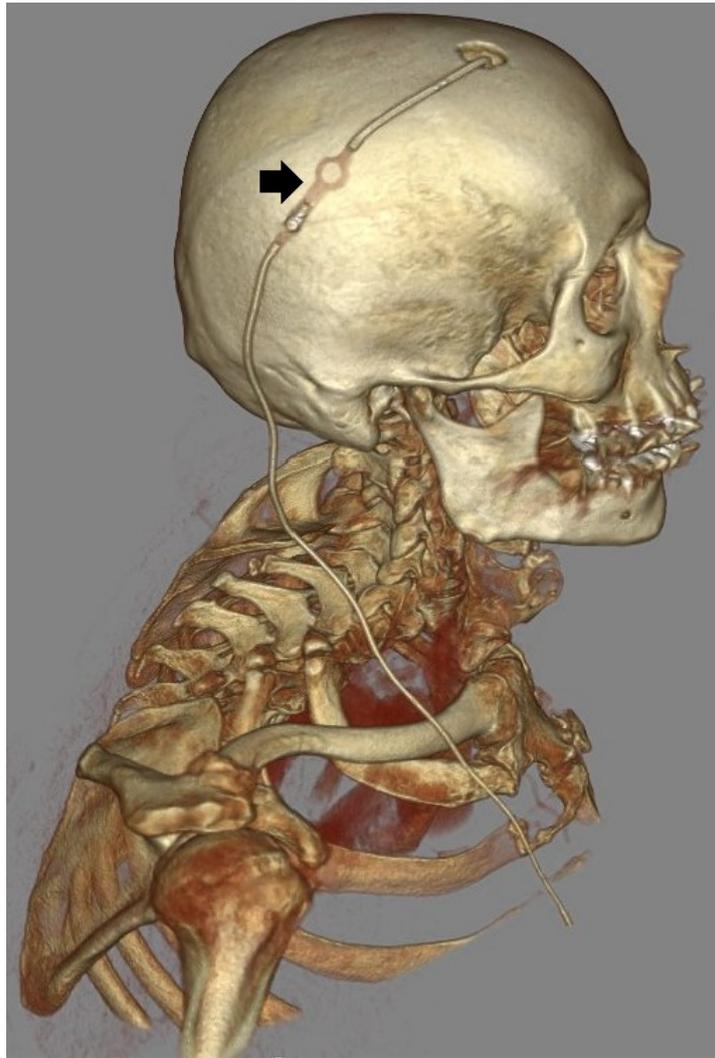
1.6) Neurochirurgische Therapieoptionen

Bei den neurochirurgischen Therapieoptionen des kindlichen Hydrozephalus werden temporäre von permanenten Ansätzen unterschieden.

Temporäre Methoden kommen insbesondere beim posthämorrhagischen Hydrozephalus nach germinaler Matrixblutung im Rahmen von Frühgeburtlichkeit zum Einsatz, wenn bei einem Kind mit einem Körpergewicht <1.500 bis 2.000 g eine therapiepflichtige Liquorzirkulationsstörung eintritt.⁴¹ Die etablierten Operationen sind in diesem Fall die Implantation ventrikulärer Reservoire, welche transkutan punktiert und aspiriert werden können, sowie ventrikulo-subgaleale Shunts, welche den Liquor aus den Ventrikeln in eine während der Operation formierte subgaleale Tasche ableiten.⁴² Externe Ventrikeldrainagen werden im Neugeborenen- und Säuglingsalter tendenziell weniger eingesetzt, stellen aber bei älteren Kindern eine sehr effektive temporäre Therapiemöglichkeit für Notfallsituationen, wie zum Beispiel einem dekompensierten akuten Okklusivhydrozephalus, sowie für Shuntinfektionen dar, wenn das infizierte permanente Shuntsystem explantiert werden muss.

Zu den permanenten Therapieoptionen zählen Liquorshunts sowie endoskopische Eingriffe, insbesondere die Drittventrikulostomie (*endoscopic third ventriculostomy*, ETV).

Liquorshunts werden in der modernen Neurochirurgie als ventrikuloperitoneale (Grafik 4) oder seltener als ventrikuloatriale Shunts implantiert.



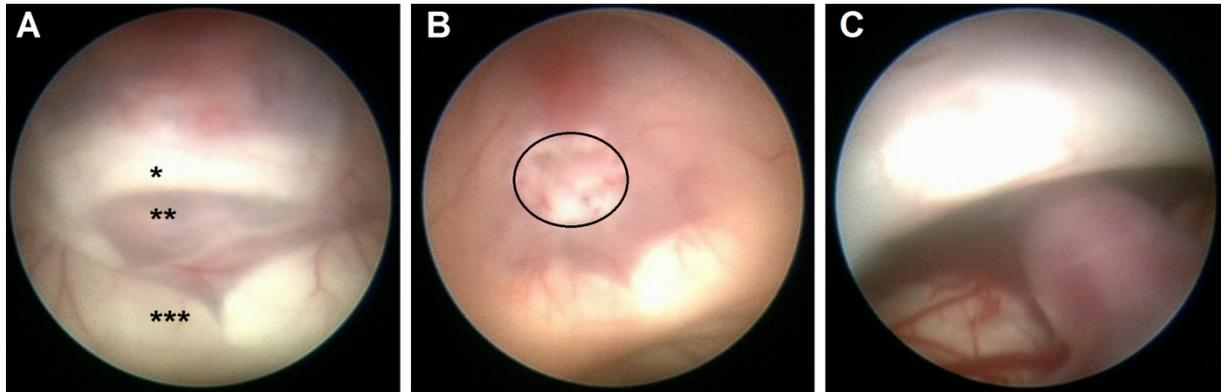
Grafik 4. Ventrikuloperitonealer Shunt *in situ*. Erkennbar sind von proximal nach distal zunächst das rechts frontale Bohrloch und der aus dem Bohrloch austretende Ventrikelkatheter, das Ventil (Pfeil) mit Reservoir und Ventilkörper sowie der distale Katheter, welcher sich bis nach abdominell fortsetzt.

Die erste ventrikuloperitoneale Ableitung wurde 1905 von Kausch durchgeführt.⁴³ Vier Jahre später gelang McClure die erste Ableitung in die großen Venen des Halses, allerdings wurde das Problem des Refluxes von venösem Blut in den Katheter erst 1988 von Xue suffizient gelöst. Die ersten Ventile zur Regulation des Liquorflusses wurden in den 1950er Jahren u.a. von Nulsen, Spitz, Pudenz, Heyer und Holter entwickelt und implantiert. Das erste kommerziell verbreitete Ventil mit transkutaner Verstellmöglichkeit wurde von Hakim konzipiert und ist seit 1989 auf dem Markt. Schätzungsweise wurden über 300 verschiedene Ventil-Typen entwickelt, welche grundsätzlich folgenden Konstruktionsprinzipien folgen: proximale

oder distale Schlitzventile, Kugelventile, Membranventile, Schwerkraftventile sowie Verstellventile.^{1,43} Allerdings ergab eine 2014 durchgeführte systematische Analyse von 22 Publikationen, welche verschiedene Ventilsysteme verglichen haben, eine fehlende Evidenz für die Nutzung eines bestimmten Systems.⁴⁴ Unter diesen Arbeiten fand sich lediglich eine prospektiv randomisierte Studie, welche die Kriterien von Klasse I Evidenz erfüllte: *The Shunt Design Trial* verglich ein Standard Differentialdruck-Ventil mit dem antisiphon Delta-Ventil (PS Medical-Medtronic, Goleta, USA) und dem autoregulierenden Sigma-Ventil (NMT Cordis, Miami, USA).⁴⁵ Weder in Bezug auf den primären Endpunkt der Studie noch im Langzeit-*follow up* ergaben sich signifikante Unterschiede zwischen den untersuchten Ventilen.^{45,46} Wissenschaftlich belegt sind hingegen Unterschiede zwischen verschiedenen Arten von zentralen und abdominellen Shuntkathetern. Antibiotika-beschichtete Katheter, welche für ca. 60 bis 130 Tage Rifampin und Clindamycin freisetzen, zeigen eine Reduktion von postoperativen Shuntinfektionen und werden daher von einer 2014 publizierten Meta-Analyse für die Nutzung bei kindlichen Shuntimplantationen empfohlen.⁴⁷ Neben der eigentlichen *hardware* eines Shunts wurden zahlreiche Studien zur Verbesserung der Präzision insbesondere bei der Implantation des ventrikulären Katheters durchgeführt.⁴⁸⁻⁵² Trotz der vermeintlich intuitiven Annahme, dass erhöhte Präzision die Shuntversagens- bzw. Komplikationsrate senken würde, können nach Kriterien der evidenzbasierten Medizin die Neuronavigation und der intraoperative Ultraschall allenfalls als Option mit unsicherer klinischer Relevanz angesehen werden; endoskopisch gestützte Ventrikelkatheteranlagen scheinen hingegen keinerlei Nutzen zu haben.⁵³

Das wichtigste endoskopische Therapieverfahren des Hydrozephalus ist die ETV. Diese wurde 1922 von Dandy zunächst als offene Operation mit Kraniotomie vorgeschlagen und 1923 von Mixer und 1936 von Scarff und Stokey weiter

ausgearbeitet.^{54,55} Bei dieser Operation wird der Boden des dritten Ventrikels anterior der *Corpora mamillaria* und posterior des *Dorsum sellae* perforiert, um so eine Verbindung zwischen den inneren Liquorräumen und dem präpontinen Subarachnoidalraum herzustellen (Grafik 5).



Grafik 5. Schrittweiser Ablauf einer ETV bei einer 16 Jahre alten Patientin mit triventrikulärem Hydrozephalus (MRT in Grafik 3): Die anatomischen Leitstrukturen (A) sind *Dorsum sellae* (*), Boden des dritten Ventrikels (**) sowie *Corpora mamillaria* (***). Stoma am Boden des dritten Ventrikels (B). Blick durch das Stoma auf die *Arteria basilaris*, das sogenannte *naked basilar artery sign* als Prädiktor des technischen Erfolgs der ETV (C).

Diese Methode trat mit der rasanten Entwicklung der Shuntsysteme zunächst in den Hintergrund, gewann aber schließlich in der zweiten Hälfte des 20. Jahrhunderts wieder an Bedeutung, als man die Nachteile einer Shuntabhängigkeit, das heißt einer Abhängigkeit von künstlichen Implantaten, erkannte. Rekate forderte schließlich im Jahr 2004: „*Every shunt failure or infection should be viewed as an opportunity to explore the possibility that the patient could become shunt independent.*“⁵⁶ Das Alter des Kindes zum Zeitpunkt der Operation sowie die Ätiologie des Hydrozephalus wurden als wichtigste Einflussgrößen für die Erfolgswahrscheinlichkeit einer ETV und damit als Hauptkriterien für die chirurgische Entscheidung des Therapieverfahrens identifiziert.⁵⁷ Klassische Indikation für eine ETV ist der triventrikuläre Hydrozephalus bei Stenose oder Verschluss des Aquäduktes bei einem Kind jenseits der Neugeborenen- und Säuglingszeit.⁵⁸

1.7) Komplikationen und Outcome

Die verlässlichsten grundlegenden Kennzahlen zu Komplikationen von Liquorshunts stammen aus prospektiv erhobenen Daten: 40% der Shunts versagen innerhalb des ersten Jahres nach Implantation und bei 10% der Patienten tritt eine Shuntinfektion auf.⁴⁵ Eine genauere Analyse des zeitlichen Verlaufs zeigt, dass 14% aller Shunts innerhalb von 30 Tagen und 24% innerhalb von 90 Tagen nach Implantation versagen.^{59,60} Ein Drittel aller Shuntrevisionen gelten als potentiell vermeidbar, da sie aufgrund von theoretisch beeinflussbaren Faktoren entstehen, zu welchen Infektionen, Fehllagen und Diskonnektionen gezählt werden.⁶¹ Frühgeborene stellen insgesamt eine Hochrisikogruppe dar, bei welcher mit einer Shuntrevisionsrate von 60% und einer Infektionsrate von 30% gerechnet werden muss.⁶²

In Abhängigkeit von der Ätiologie des Hydrozephalus weisen 12 bis 50% der betroffenen Kinder kognitive Einschränkungen auf.⁶³ Schulische Probleme zeigen sich bei 20 bis 60% der Kinder.⁶³ Im Erwachsenenalter haben 40 bis 60% der als Kind mit einem Shunt versorgten Patienten eine normale schulische Ausbildung erhalten und 34 bis 42% arbeiten im kompetitiven Arbeitsmarkt.^{63,64} Bezüglich des sozialen Lebens ist bekannt, dass 67% der Patienten im Haushalt der Eltern verbleiben, 10% alleine leben und 6% mit einem Partner zusammen leben.⁶⁵

Ähnlich wie bei Shuntoperationen ist auch die ETV eine neurochirurgische Operation mit geringer Morbidität und Mortalität in der intra- und perioperativen Phase. Die Mortalität der ETV wird mit unter 1% angegeben und die transiente sowie permanente Morbidität liegen bei 1% bis 8% bzw. 0,5 bis 2%.^{66,67} Ursache von Mortalität ist meist eine Subarachnoidalblutung bei Verletzung der *Arteria basilaris*

oder ihrer Äste, die Morbidität umfasst hauptsächlich Liquorfisteln, Augenkoordinationsstörungen oder hormonelle Störungen.

Die Lebensqualität im Langzeitverlauf scheint sich zwischen Shunt und ETV nicht zu unterscheiden.⁶⁸

Allerdings bestehen Unterschiede in der primären Erfolgswahrscheinlichkeit einer ETV: Die Analyse des Einflusses insbesondere von Lebensalter und Ätiologie des Hydrozephalus führte zur Entwicklung und weiteren Validierung des *ETV Success Scores*.^{69,70} Hauptvorteil der ETV gegenüber einem Shunt sind die geringen Langzeitprobleme. So ist bei optimalen Kandidaten des *ETV Success Scores* bereits früh nach Operation das Komplikationsrisiko geringer als bei Shuntanlage und auch bei weniger optimalen Kandidaten ist nach 3 bis 6 Monaten das Risiko einer Komplikation der ETV geringer als das Risiko einer Shuntkomplikation.⁶⁸ Große Aufmerksamkeit wird hydrozephalen Kindern in den ersten beiden Lebensjahren zuteil, welche zwar die längste Zeit mit ihrer Liquorableitung leben müssen und daher theoretisch besonders unter Shuntkomplikationen leiden würden, bei denen jedoch nach dem *ETV Success Scores* die geringste Erfolgswahrscheinlichkeit einer ETV besteht.⁶⁹ Kleinere Serien bestreiten dies zwar, die Hypothese wird aber durch erste Ergebnisse einer aktuellen prospektiven randomisierten Studie bestätigt, welche Shunt und ETV bei Kindern jünger als 24 Monate vergleicht.^{71,72} Nach 3 Monaten liegt die Erfolgsrate eines Shunts bei 95%, einer ETV hingegen bei 68%. Die Ergebnisse der 5-Jahres-Nachbeobachtung der *International Infant Hydrocephalus Study* stehen jedoch noch aus.⁷² Je nach finalen Ergebnissen dieser Studie könnte der Versuch einer primären ETV auch in dieser Altersgruppe angesichts der geringen direkten operativen Komplikationen gerechtfertigt sein, denn ein sekundärer Shunt

bei fehlgeschlagener primärer endoskopischer Therapie hat keine höhere Komplikationsrate als ein primärer Shunt.⁷³

Im umgekehrten Fall jedoch, das heißt bei Vorhandensein eines Shunts, reduziert sich die Wahrscheinlichkeit einer erfolgreichen sekundären ETV um ca. 10%.⁶⁹

1.8) Fragestellungen und Ziele der eigenen Arbeiten

- a) Lässt sich die Präzision der Platzierung des ventrikulären Katheters durch Nutzung des intraoperativen Ultraschalls verbessern?
- b) Lässt sich durch verbesserte Lage des ventrikulären Katheters die Shundysfunktionsrate reduzieren?
- c) Welche Einfluss haben verschiedene Shuntventile auf das Outcome?
- d) Lassen sich in Abhängigkeit vom implantierten Shuntventil, insbesondere Differentialdruckventil versus Gravitationsventil, im klinischen Alltag Unterschiede in der Überdrainage-Rate detektieren?
- e) Wie lässt sich Morbidität und Mortalität in der pädiatrischen Neurochirurgie erfassen und vergleichen und welchen Stellenwert haben liquorassoziierte Eingriffe?
- f) Wie lässt sich die Qualität einer Shuntoperation messen und vergleichen?
- g) Zielsetzung war zudem das Etablieren eines neuen patientenzentrierten und potentielle Verbesserungsoptionen eröffnenden Qualitätsindikators, der *Negative Shunt Revision Rate*.

- h) Welchen Einfluss hat die Anwesenheit eines Shunts bei heranwachsenden Menschen im Alltag in Bezug auf Symptome, insbesondere Kopfschmerzen, sowie Einschränkungen?

2) Eigene Arbeiten

2.1) Real-time ultrasound guidance for ventricular catheter placement in pediatric cerebrospinal fluid shunts.

Beez T, Sarikaya-Seiwert S, Steiger HJ, Hänggi D

Childs Nerv Syst. 2015 Feb;31(2):235-41

DOI: 10.1007/s00381-014-2611-4

PMID: 25564197

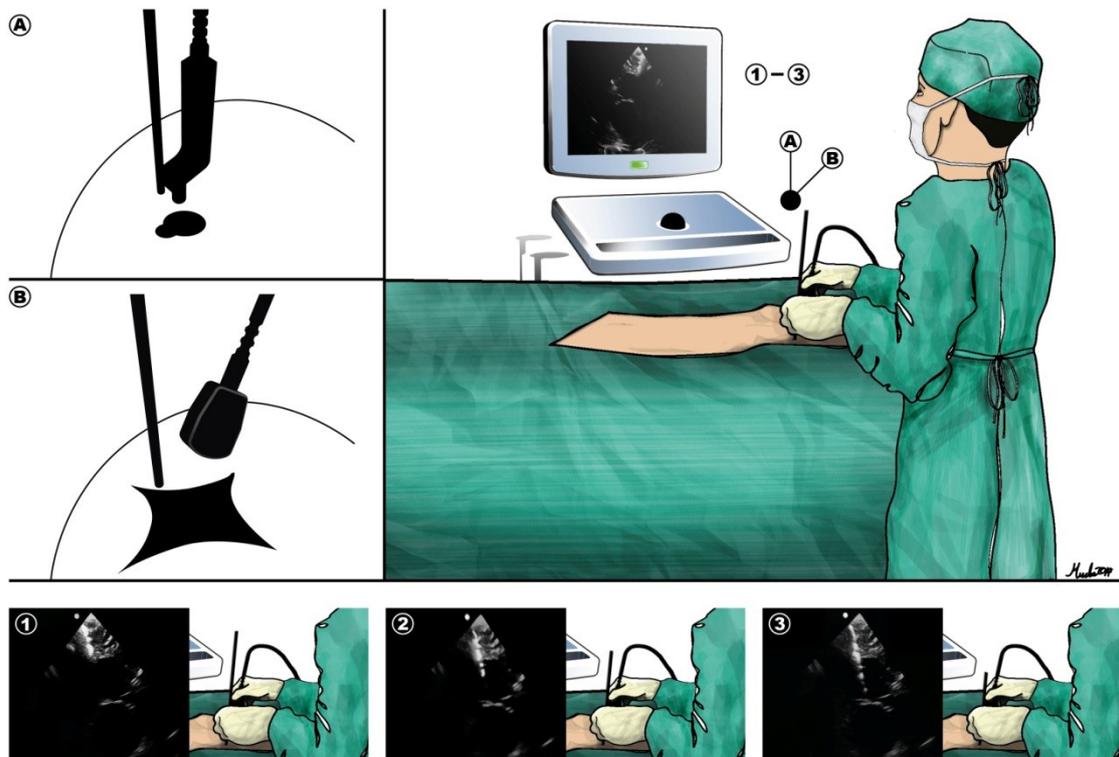
Einleitung:

Das Versagen eines Liquorshunts führt zu zusätzlichen Krankenhausaufenthalten und Operationen mit potentieller Morbidität und selten sogar Mortalität für den Patienten, Belastung der betroffenen Familien sowie zusätzlichen Kosten für das Gesundheitssystem.^{63,74} Die Reduktion der Shuntversagensrate stellt somit einen wichtigen Aspekt bei der Behandlung des kindlichen Hydrozephalus dar. Einige Arbeiten weisen darauf hin, dass eine optimale Platzierung des ventrikulären Katheters einen Einfluss auf das Shuntüberleben haben könnte.^{75,76} Die klassische chirurgische Vorgehensweise beim Implantieren eines Ventrikelkatheters ist die auf anatomischen Landmarken basierende Freihandpunktion. Diese Technik führt jedoch in bis zu 40% der Fälle zu einer suboptimalen Lage des Katheters.⁷⁷ Ziel unserer Studie war die Analyse des Einflusses von intraoperativem Ultraschall auf die Präzision der Ventrikelkatheterplatzierung sowie auf das Shuntüberleben bei Kindern.

Methoden:

Nach Genehmigung durch die lokale Ethikkommission (Studiennummer 4588) wurden alle Kinder retrospektiv identifiziert, welche zwischen April 2012 und März 2013 einen ventrikuloperitonealen Shunt unter Einsatz des intraoperativen Ultraschalls erhalten haben. Als Kontrollgruppe diente eine nach Alter gematchte Kohorte mit Freihandpunktion. Die Größe des Ventrikelsystems wurde mittels der *frontal occipital horn ratio* (FOHR) quantifiziert.⁷⁸ Die Position des Ventrikelkatheters in der postoperativen Bildgebung wurde nach folgendem Schema bewertet: Grad 1 entspricht einer optimalen Position im Ventrikel mit Liquor zu allen Seiten des Katheters, Grad 2 entspricht allen anderen Lagen im Ventrikel, d.h. kontralateral oder mit Kontakt zu Septum oder Ventrikelwänden und Grad 3 beschreibt einen fehlplatzierten Katheter außerhalb des Ventrikelsystems.⁵⁰

Für die Eingriffe wurde ein Ultraschallgerät (ProSound Alpha 6, Hitachi Aloka Medical Ltd., Tokyo, Japan) mit Bohrlochsonde (3.0–7.5 MHz, UST-52114P, Hitachi Aloka Medical Ltd., Tokyo, Japan) bzw. konvexer Sonde (3.75–10 MHz, UST-9120, Hitachi Aloka Medical Ltd., Tokyo, Japan) genutzt. Bei Kindern mit geschlossener Fontanelle erfolgte nach Anlage eines typischen Bohrloches mit einem Durchmesser von 14mm eine kleine laterale Erweiterung um 3-5mm mittels Stanze, wo die Dura inzidiert wurde. Die Bohrlochsonde wurde so eingeführt, dass der Ventrikelkatheter durch den Führungskanal der Sonde und über die laterale Erweiterung des Bohrloches vorgeschoben werden konnte. Bei Kindern mit offener anteriorer Fontanelle wurde die konvexe Sonde genutzt, um transfontanellär zu sonografieren. In beiden Fällen wurde das Einführen des Katheters vom Cortex bis hin zur Endposition im Ventrikel kontinuierlich in Echtzeit visualisiert, um die Trajektorie bei Bedarf zu korrigieren (Grafik 6).



Grafik 6. Illustration des operativen Vorgehens bei der ultraschallgesteuerten Ventrikelkatheteranlage mit Bohrlochsonde (A) oder konvexem Schallkopf (B). Die Darstellung des vordringenden Katheters erfolgt kontinuierlich und in Echtzeit (1-3).

Ergebnisse:

Studiengruppe (n = 17) und Kontrollgruppe (n = 14) wiesen keine signifikanten Unterschiede in den Basisparametern auf, insbesondere bezogen auf das mittlere Alter (4,7 und 4,3 Jahre), die präoperative FOHR (0,45 und 0,43) und die mittlere Dauer der Operation (51 Minuten und 49 Minuten). In der Studiengruppe konnte eine Katheterposition entsprechend Grad 1 in 6 Fällen (35 %) und Grad 2 in 11 Fällen (65 %) erreicht werden. Eine Grad 3 Lage fand sich in dieser Gruppe hingegen nicht. In der Kontrollgruppe zeigte sich eine Lage entsprechend Grad 1 bei 2 Patienten (18 %), Grad 2 bei 3 Patienten (27 %) und Grad 3 bei 9 Patienten (43 %). Der Unterschied zwischen beiden Gruppen bezüglich der Katheterlage erreichte

statistische Signifikanz ($P = 0,0029$). Die Shuntversagensrate war am höchsten bei Kathetern mit Grad 3 Lage (83 %).

Diskussion:

Diese Studie zeigte eine signifikante Verbesserung der Lage des ventrikulären Katheters bei Shuntimplantationen im Kindesalter. Insbesondere konnten bei Einsatz des intraoperativen Ultraschalls Fehllagen, welche mit dem höchsten Risiko einer späteren Shuntfehlfunktion einhergehen, komplett vermieden werden.

Ähnliche Ergebnisse konnten auch mit anderen intraoperativen Methoden zur Erhöhung der Präzision erzielt werden, wie z.B. Neuronavigation oder Kombination aus präoperativer Trajektorienplanung und speziellen Führungsinstrumenten.^{49–}

^{51,79,80} Im Gegensatz dazu scheint jedoch die Platzierung des Ventrikelkatheters unter endoskopischer Kontrolle nicht mit einer Verbesserung der Präzision einherzugehen.⁵² Neben einer Insuffizienz der Methode selbst könnte eine Verlagerung der relativ weichen Silikon-Katheter innerhalb der Ventrikel im postoperativen Verlauf eine Ursache sein. Dieses Phänomen wurde allerdings auch für ultraschallgesteuerte Katheterimplantationen beschrieben und scheint besonders bei Kindern jünger als 6 Monate mit dünnem kortikalen Mantel bzw. sehr großen Ventrikeln aufzutreten.⁸¹

Zusammenfassend wurden in einer 2014 publizierten Meta-Analyse sowohl intraoperativer Ultraschall als auch Neuronavigation als Optionen für die routinemäßige Nutzung bei Shuntimplantationen im Kindesalter empfohlen.⁵³

Schlussfolgerung:

Die Nutzung des intraoperativen Ultraschalls verbessert die Präzision der Ventrikelkatheterplatzierung und vermeidet Fehllagen, welche einen Risikofaktor für Shuntversagen darstellen. Da bei der beschriebenen Methode keine zusätzliche Belastung des Patienten entsteht, z.B. im Sinne einer Strahlenexposition oder längeren Operationszeit, sollte die Katheterplatzierung unter Ultraschallkontrolle der Freihandpunktion vorgezogen werden.

2.2) Role of ventriculoperitoneal shunt valve design in the treatment of pediatric hydrocephalus--a single center study of valve performance in the clinical setting.

Beez T, Sarikaya-Seiwert S, Bellstädt L, Mühmer M, Steiger HJ

Childs Nerv Syst. 2014 Feb;30(2):293-7

DOI: 10.1007/s00381-013-2244-z

PMID: 23900632

Einleitung:

Hinsichtlich der Wahl des Ventils bei pädiatrischen Shunts kann auf Basis von publizierten Studien keine evidenzbasierte Empfehlung gegeben werden, da bisher keine Überlegenheit eines bestimmten Systems gezeigt werden konnte.⁴⁴ Die qualitativ hochwertigste Studie zu diesem Thema war *The Shunt Design Trial*.⁴⁵ Diese prospektive randomisierte Studie konnte jedoch ebenfalls keinen Unterschied zwischen den verglichenen Ventilen verschiedener Bauart identifizieren.

Ein weiterer Aspekt bei der Wahl des Ventils, insbesondere angesichts formal fehlender oder zumindest nicht wissenschaftlich belegter Vorteile, sind die Kosten. Verstellbare Ventile kosten mehr als doppelt so viel wie nicht verstellbare Ventile.⁸² Noch dramatischer ist der Preisunterschied zwischen dem in Indien gefertigten Chhabra-Ventil (Surgiwear LTD, Shahjahanpur, Indien) für 35 US Dollar und dem in den USA hergestellten Codman Hakim Micro Precision-Ventil (Codman & Shurtleff, Inc., Raynham, USA) für 650 US Dollar, welche in einer klinischen Studie identische Ergebnisse lieferten.⁸³ Zudem fanden sich auch *in vitro* keine signifikanten Unterschiede in der Funktion des Chhabra-Ventils und teurerer Ventile aus westlicher Produktion.⁸⁴

An unserer Institution hat sich bei fehlender Datenlage historisch ein auf Erfahrung basiertes Protokoll für die Wahl des Shuntventils ergeben: Bei Früh- oder Neugeborenen, Säuglingen und Kleinkindern wird das nicht verstellbare Gravitationsventil paediGAV (Aesculap AG, Tuttlingen, Deutschland) aufgrund der kleinen Bauart und der propagierten Vermeidung einer Überdrainage eingesetzt. Bei älteren Kindern wird das größere, aber verstellbare Differentialdruckventil Codman Hakim (Codman & Shurtleff, Inc., Raynham, USA) genutzt.

Ziel der vorliegenden Studie war der Vergleich dieser beiden Ventile in Bezug auf Shuntüberleben und Komplikationsrate im klinischen Alltag.

Methoden:

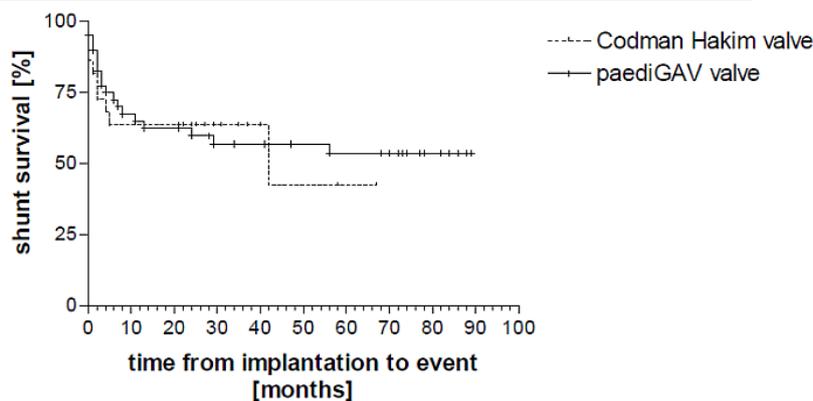
Patienten mit einem Alter von 16 Jahren oder jünger, welche an unserer Institution zwischen Januar 2005 und Dezember 2010 einen Shunt erhalten haben, wurden retrospektiv identifiziert. Folgende Daten wurden aus der elektronischen Patientendokumentation extrahiert: Ätiologie des Hydrozephalus, Alter bei Shuntimplantation, Ventiltyp, Zeit bis zum Shuntversagen sowie Ursache des Shuntversagens. Neben deskriptiver Statistik erfolgte eine Analyse mittels Mann-Whitney-Test sowie Kaplan-Meier-Methode.

Ergebnisse:

Die paediGAV-Gruppe umfasste 44 und die Codman Hakim-Gruppe 29 Patienten. Die mittlere Beobachtungsdauer lag bei 34 Monaten. Ein Patient verstarb an einer neonatalen Sepsis und drei Patienten an Hirntumoren. Es traten keine shunt-assoziierten Todesfälle auf. Erwartungsgemäß spiegelte sich das institutionelle Protokoll in den Basisdaten beider Gruppen wider: Das mittlere Alter bei Shuntanlage lag in der paediGAV-Gruppe bei 18 Monaten (0 bis 154 Monate) und in der Codman

Hakim-Gruppe bei 70 Monaten (1 bis 180 Monate). Zugrundeliegende Hauptursachen des Hydrozephalus waren Blutung und Fehlbildung in der erstgenannten Gruppe sowie Tumor und Fehlbildung in der zweitgenannten Gruppe. In der paediGAV Gruppe erreichten 53% der Patienten den Endpunkt des Shuntversagens nach einer mittleren Dauer von 7 Monaten (Grafik 7). In der Codman Hakim Gruppe traf dies auf 56% zu und die mittlere Dauer lag bei 8 Monaten.

Valve type	No. of patients	No. of pat. with shunt failure	Shunt failure rate
paediGAV	40	21	53%
Codman Hakim	25	14	56%



Grafik 7. Vergleich des Shuntüberlebens von paediGAV-Gruppe und Codman Hakim-Gruppe.

Hinsichtlich der Ursachen des Shuntversagens zeigten sich keine signifikanten Unterschiede, insbesondere nicht in Hinblick auf Über- oder Unterdrainage. Häufigste Ursache waren mit 64 bzw. 59% aller Shunt Dysfunktionen jeweils mechanische Probleme. Die stratifizierte Analyse der Gesamtkohorte identifizierte junges Alter bei Implantation (<12 Monate) sowie posthämorrhagischen Hydrozephalus als Risikofaktoren für Shuntversagen.

Diskussion:

Diese Studie zeigte keine signifikanten Unterschiede zwischen den beiden untersuchten Ventilen bei der klinischen Anwendung im Rahmen eines

institutionellen Protokolls, welches im Wesentlichen auf dem Alter des Kindes basiert. Insbesondere fand sich kein Unterschied bezüglich des Risikos für Shuntversagen sowie Überdrainage. Das höchste Risiko fand sich bei Implantation eines Shunts im ersten Lebensjahr sowie bei posthämorrhagischem Hydrozephalus.

Überdrainage stellte sich nicht als wesentliches klinisches Problem dar. Die Infektionsrate lag im Bereich der Literatur, wobei anzumerken ist, dass alle in dieser Studie eingeschlossenen Eingriffe nach einem an die publizierten Vorgaben angepassten Protokoll zur Infektionsreduktion durchgeführt worden sind.⁸⁵⁻⁸⁷ Hauptursache für Shuntfehlfunktionen waren hingegen mechanische Probleme, welche hauptsächlich Okklusionen des zentralen Katheters sowie des Ventils umfassen.

Schlussfolgerung:

Diese Studie bestätigt die Ergebnisse vorheriger Publikationen, welche ebenfalls keine signifikanten Unterschiede zwischen verschiedenen Ventilen beschreiben.⁴⁴ Eine Änderung unseres internen klinischen Protokolls ist auf Basis dieser Erkenntnisse nicht erforderlich gewesen.

2.3) Prospective review of 30-day morbidity and mortality in a paediatric neurosurgical unit.

Campbell E, Beez T, Todd L

Childs Nerv Syst. 2017 Mar;33(3):483-489

DOI: 10.1007/s00381-017-3358-5

PMID: 28247111

Einleitung:

„Tout chirurgien porte en lui un petit cimetière dans lequel il va de temps en temps faire oraison. Cimetière d’amertume et d’hysope, auquel il demande la raison de certains de ses succès.“

Henri Marie René Leriche⁸⁸

In den vergangenen Jahren hat das Interesse vieler Seiten an den Komplikationen eines Chirurgen, also in Leriche's Worten an seinem persönlichen „Friedhof“, deutlich zugenommen. Zum Beispiel wird in Großbritannien seit 2014 für einige chirurgische Disziplinen eine Veröffentlichung von Komplikations- und Mortalitätsraten für einzelne Institutionen und sogar individuelle Chirurgen (Fachärzte) gesetzlich vorgeschrieben. Dieses Vorgehen wirft auf verschiedenen Ebenen Fragen auf: Welche Daten können zur Berechnung solcher Qualitätsmarker genutzt werden? Sind diese Qualitätsmarker überhaupt sachdienlich, d.h. liefern sie für den Patienten nützliche Information, sind sie reproduzierbar, allgemeingültig sowie vergleichbar und erlauben sie letztendlich eine Überwachung und Verbesserung der Qualität? Welche Auswirkungen hat die Veröffentlichung auf den einzelnen Chirurgen und sollte man in diesem Licht als Teil der „Optimierung“ seiner Qualitätsmarker auf chirurgische Ausbildung oder Durchführung von Hochrisiko-Operationen verzichten?^{89,90}

Während die letzte Frage der juristischen und ethischen Debatte überlassen werden sollte, haben wir uns im Rahmen der vorliegenden Studie den ersten beiden Fragen gewidmet. Ziel der Studie war die Entwicklung und Implementierung eines auf die pädiatrische Neurochirurgie zugeschnittenen Systems zur Erfassung von Morbidität und Mortalität bei neurochirurgischen Patienten jünger als 16 Lebensjahren.

Methoden:

Das genutzte System basiert auf einem zuvor von Drake et al. publizierten Modell, welches kritische Ereignisse unter den Oberbegriffen „chirurgische“ oder „medizinische“ Komplikation in verschiedene Kategorien klassifiziert (Tabelle 2).⁹¹

Chirurgische Komplikationen	Neurologisches Defizit (neu aufgetreten) Meningitis (außer bei externer Liquordrainage) Krampfanfall Wundinfektion (Verdacht oder Nachweis) Liquorfistel Infektion bei externer Ventrikel- oder Lumbaldrainage Shuntokklusion Shuntinfektion (Verdacht oder Nachweis) Postoperative Einblutung Konservative Therapie Postoperative Einblutung Revisionsoperation Postoperative Infarzierung Andere
Medizinische Komplikationen	Kardial Respiratorisch (inklusive Pneumonie) Gastrointestinal und hepatisch Renal und ableitende Harnwege Hämatologisch und thromboembolisch Metabolisch Kognitiv Andere

Tabelle 2. Klassifikation perioperativer Komplikationen nach Drake et al.⁹¹

Zusätzlich wurde der Schweregrad des Ereignisses entsprechend den 5 Kategorien der *NHS Greater Glasgow & Clyde Incident Management Policy* in „negligible“ bis „extreme“ eingeteilt.⁹² Eine aufgrund einer Komplikation notwendige ungeplante Folge-Operation bedingt mindestens eine Kategorie 3. Ein kritisches Ereignis wurde ab einem Wert von gleich oder größer als Kategorie 3 als „signifikant“ bezeichnet.

Grad	Bezeichnung	Beschreibung
1	Unerheblich	Reduzierte Qualität des Patientenkomfort bzw. des Ergebnisses ohne direkten Zusammenhang zur medizinischen Versorgung
2	Geringgradig	Verschlechterung des Ergebnisses der med. Versorgung – sofort behebbar
3	Mittelgradig	Verschlechterung des Ergebnisses der med. Versorgung – Erholungszeit <1 Woche
4	Hochgradig	Verschlechterung des Ergebnisses der med. Versorgung – Erholungszeit >1 Woche
5	Extrem	Verschlechterung des Ergebnisses der med. Versorgung – Langzeitschäden

Tabelle 3. Klassifikation des Schweregrades kritischer Ereignisse nach der *NHS Greater Glasgow & Clyde Incident Management Policy*.⁹²

Über einen Zeitraum von 2 Jahren wurden die Daten prospektiv gesammelt. Konkret wurden alle operativen Fälle in wöchentlichen Team-Besprechungen diskutiert und bei Vorliegen eines kritischen Ereignisses entsprechend des oben genannten Systems klassifiziert. Alle kritischen Ereignisse innerhalb von 30 Tagen nach einer Index-Operation wurden registriert. Mehrere Ereignisse eines Patienten innerhalb von 30 Tagen wurden jeweils als separate Komplikation gezählt.

Ergebnisse:

Zwischen Januar 2014 und Dezember 2015 wurden 549 Operationen bei 287 Patienten durchgeführt, wovon 124 weiblich und 163 männlich waren und das mittlere Alter bei 5,8 Jahren lag. 25% der Eingriffe wurden bei Kindern im ersten

Lebensjahr durchgeführt. 75% der Operationen waren dringliche oder notfallmäßige Eingriffe und 25% waren elektiv geplante Operationen. Während dieses Zeitraumes sind 6 Kinder innerhalb von 30 Tagen nach Operationen verstorben, was eine Mortalitätsrate von 2,1% ergibt. Es wurden 190 kritische Ereignisse registriert, wovon 130 als signifikante Ereignisse klassifiziert worden sind. Die Rate an signifikanten kritischen Ereignissen (*significant adverse event rate*) lag somit bei 20%. Von den kritischen Ereignissen waren 159 chirurgische und 31 medizinische Komplikationen. Das häufigste kritische Ereignis insgesamt war eine postoperative Liquorfistel und das häufigste signifikante kritische Ereignis war eine Shunt Dysfunktion durch Okklusion. Operationen mit Liquorableitung aller Art (Shunts, externe Ventrikeldrainage, ventrikuläre endoskopische Operationen) hatten eine signifikant höhere Rate an signifikanten kritischen Ereignissen von 26,5% im Vergleich zu anderen kraniellen (12,2%) oder spinalen (21,9%) Eingriffen ($p < 0,001$). Die Rate an ungeplanten Folge-Operationen (*unscheduled return to theatre rate*) lag bei 15% aller Index-Operationen und die Infektionsrate (*surgical site infection rate*) bei 5,5%.

Diskussion:

Zusammenfassend ließ sich das von uns entwickelte Modell ohne nennenswerte Probleme in die Arbeitsabläufe einer Abteilung für pädiatrische Neurochirurgie integrieren. Wir konnten anhand der prospektiven gewonnenen Daten sowohl individuelle Erkrankungen bzw. Operationen detailliert analysieren (z.B. liquorableitende Eingriffe) als auch in Literatur gebräuchliche Kennzahlen oder Qualitätsindikatoren zum Vergleich berechnen.

Die von uns festgestellte *significant adverse event rate* von 20% steht in Übereinstimmung mit den bisher publizierten Erkenntnissen für die pädiatrische Neurochirurgie: Bei Drake et al. lag diese Rate bei 16,4% und bei van Lindert et al.

bei 20,2%.^{91,93} Unsere *unscheduled return to theatre rate* von 15% bewegt sich ebenfalls im Bereich des publizierten Spektrums von 10,5% bis 17%.^{93,94} Dies trifft ebenso auf unsere *surgical site infection rate* von 5,5% zu, da sich diesbezüglich je nach analysiertem neurochirurgischem Patientenkollektiv Infektionsraten von 3,3% bis 15,4% finden.^{93,95}

Im Gegensatz dazu erkannten wir, dass die *significant adverse event rate* für liquorableitende Operationen in unserem Kollektiv mit 26,5% über den Werten aus der Literatur liegt, welche mit 15,4% bis 21,8% angegeben werden.^{91,93} Eine daraufhin durchgeführte ergänzende Analyse dieser Subgruppe ergab, dass die Mehrzahl dieser Fälle auf Shuntversagen infolge einer Okklusion binnen 30 Tagen nach Index-Operation entfällt. Es wurden deshalb nach diesbezüglicher Literatur-Recherche und Abgleich mit den internen Vorgehensweisen Bereiche mit Verbesserungspotential identifiziert und entsprechende Maßnahmen implementiert, welche seither mit dem publizierten Modell hinsichtlich ihres positiven Einflusses analysiert werden. Diese Ergebnisse stehen derzeit noch aus.

Schlussfolgerung:

Das in dieser Publikation vorgestellte Modell zur prospektiven Erfassung von Morbidität und Mortalität in der pädiatrischen Neurochirurgie ließ sich in den Arbeitsfluss der Abteilung integrieren und lieferte robuste Resultate, welche sowohl den externen Vergleich mit der Literatur als auch die interne Erkennung von Problemfeldern ermöglichen. Ob das Modell auch den Einfluss von Maßnahmen zur Qualitätsverbesserung detektieren wird, ist Gegenstand derzeit laufender Analysen.

2.4) Comparison of Quality Metrics for Pediatric Shunt Surgery and Proposal of the Negative Shunt Revision Rate.

Beez T, Steiger HJ

World Neurosurg. 2017 Oct. [Epub ahead of print]

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PMID: 28987855

Einleitung:

Die Shuntimplantation stellt eine wichtige Säule der Therapie des kindlichen Hydrozephalus dar und zählt zu den am häufigsten durchgeführten Eingriffen in der pädiatrischen Neurochirurgie. Shuntassoziierte Komplikationen sind häufig und bedeuten sowohl eine zusätzliche Belastung für den Patienten als auch eine finanzielle Belastung der Gesundheitssysteme.^{74,96,97} Es wurden daher zahlreiche Qualitätsindikatoren entwickelt, um die Ergebnisse von Shuntoperationen vergleichbar zu machen und eine Qualitätssicherung und –optimierung zu ermöglichen.

Ein Blick über den „Tellerrand“ der Neurochirurgie zeigt, dass ähnliche Bestrebungen auch in anderen chirurgischen Disziplinen bestehen: In der Abdominalchirurgie wurde die negative Appendektomie-Rate definiert als der Anteil der nicht-inzidentellen Entfernungen eines gesunden Appendix. Die Rate lag in älteren Publikationen bei ca. 15%.⁹⁸ Es konnte gezeigt werden, dass negative Appendektomien mit Morbidität und Kosten einhergehen. Durch Implementation von radiologischen und klinischen Algorithmen konnte die Rate auf bis zu 3% gesenkt werden.⁹⁹

Ziele der vorliegenden Studie waren es, die wichtigsten Qualitätsindikatoren an unserem Patientenkollektiv zu bestimmen und im Kontext der Literatur zu bewerten

sowie die *Negative Shunt Revision Rate* als neuen Indikator in Anlehnung an die negative Appendektomie-Rate zu etablieren.

Methoden:

Nach Genehmigung durch die lokale Ethikkommission (Studennummer 5949R) wurden Patienten jünger als 18 Lebensjahre identifiziert, welche im Jahr 2015 eine Implantation oder Revision eines Liquorshunts erhalten haben. Alter bei Operation, Geschlecht, Ätiologie des Hydrozephalus und Anzahl der vorherigen Shuntoperationen wurden registriert. Die Größe des Ventrikelsystems wurde mittels der FOHR quantifiziert.⁷⁸ Shuntversagen binnen 30 Tagen bzw. 90 Tagen wurde auf die Index-Operation bezogen. Die Ätiologie des Shuntversagens wurde in proximales oder distales Versagen, Ventilversagen, Infektion oder Wundrevision eingeteilt. Eine Infektion wurde definiert als klinische Diagnose (Fieber, meningitische Zeichen, Infektkonstellation im Liquor, lokale Infektionszeichen an der Haut über dem Shunt) mit oder ohne positiven Keimnachweis.

Folgende Qualitätsindikatoren wurden entsprechend den Vorgaben der Originalpublikationen an unserem Kollektiv bestimmt: *Surgical Activity Rate*, *Revision Quotient*, *30-Day Shunt Malfunction Rate*, *90-Day Global Shunt Revision Rate* und *Preventable Shunt Revision Rate*.^{100,59,61,60}

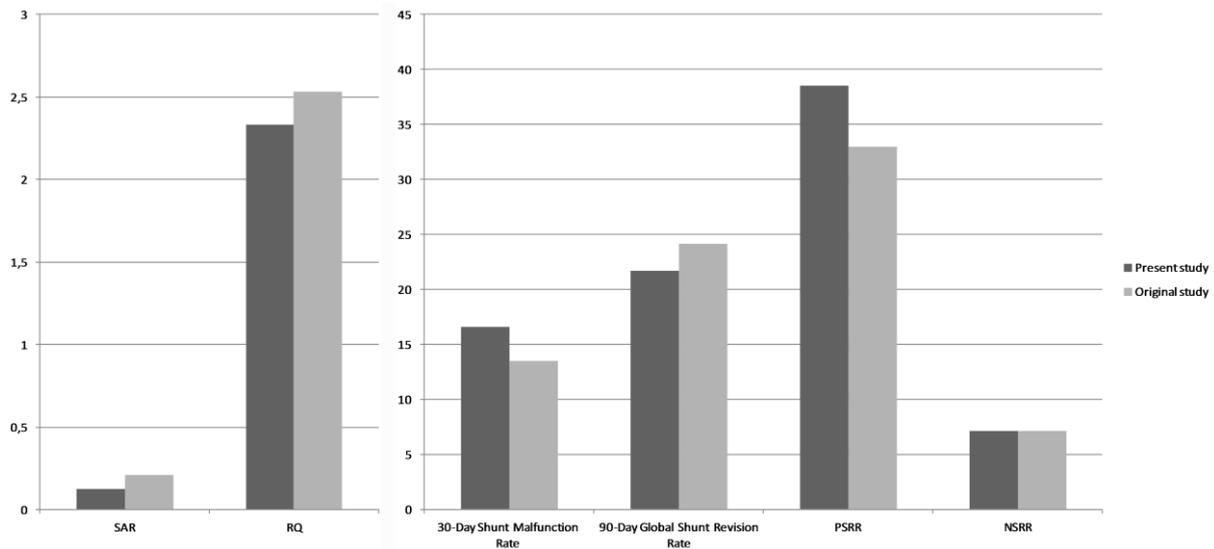
Die neue *Negative Shunt Revision Rate* wurde definiert als der Anteil aller Shuntrevisionen pro Jahr, bei welchen intraoperativ ein ordnungsgemäß funktionierendes Shuntsystem nachgewiesen wurde. Im OP-Bericht der Revision musste *per definitionem* eindeutig ein funktionierender Shunt beschrieben worden sein. Mögliche Konsequenzen waren Belassen des alten Shunts *in situ* oder

komplette oder teilweise Revision des funktionierenden Systems aus „prophylaktischen“ Gründen.

Ergebnisse:

Sechzig Shuntoperationen wurden eingeschlossen, bestehend aus 18 (39%) Neuanlagen sowie 42 (70%) Revisionen. Das Patientenkollektiv bestand aus 19 weiblichen und 20 männlichen Kindern mit einem mittleren Alter bei Operation von 18 Monaten. Die mittlere präoperative FOHR betrug 0,42 (0,32 bis 0,60). Die häufigsten Ätiologien des Hydrozephalus waren posthämorrhagisch (N = 16, 41%), kongenital (N = 11, 28%) und tumorbedingt (N = 8, 21%). Es kam in 13 Fällen zu Shuntversagen, hauptsächlich durch proximales Versagen (N = 6, 46%). Die *Surgical Activity Rate* war 0,127, der *Revision Quotient* war 2,333, die *30-Day Shunt Malfunction Rate* betrug 0,166, die *90-Day Global Shunt Revision Rate* lag bei 21,7% und die *Preventable Shunt Revision Rate* belief sich auf 38,5% (Grafik 8).

Die *Negative Shunt Revision Rate* betrug 7,1%. Es handelte sich um drei Revisionsoperationen, welche die Kriterien einer negativen Shuntrevision erfüllten. Bei allen drei Patienten lagen präoperativ klinische Verdachtsmomente vor: Bei einem Patienten bestand eine gespannte Fontanelle, bei einem weiteren Patienten zeigten sich Erbrechen und ein sich verzögert füllendes Shuntreservoir und beim dritten Kind lag lediglich eine verzögerte Füllung des Reservoirs vor. Bei allen drei Kindern war die Ventrikelweite radiologisch stabil. In zwei Fällen wurde der Shunt unverändert belassen und in einem Fall revidiert.



Grafik 8. Vergleich der erfassten Qualitätsindikatoren an unserer Kohorte mit den Referenzwerten. SAR = *Surgical Activity Rate*, RQ = *Revision Quotient*, PSRR = *Preventable Shunt Revision Rate*, NSRR = *Negative Shunt Revision Rate*.

Diskussion:

Die vorliegende Studie konnte zeigen, dass sich die publizierten Qualitätsindikatoren mit moderatem Aufwand aus verfügbaren Daten kalkulieren lassen. Für unser Kollektiv lagen diese Indikatoren im Bereich der publizierten Referenzwerte.

Qualitätsindikatoren sollen messbar, modifizierbar, sachlich sinnvoll und patientenorientiert sein.^{61,101} Während aus unserer Sicht die untersuchten Indikatoren die ersten drei Kriterien durchaus erfüllen, halten wir das letzte Kriterium für problematisch. Ein Patient wird eine Shuntrevision *per se* sehr ähnlich erleben, unabhängig davon, ob es sich nach Definition der *Preventable Shunt Revision Rate* um eine vermeidbare Ursache oder um eine andere Art der ungeplanten Revisionsoperation handelt. Allerdings reflektiert eine niedrige *Preventable Shunt Revision Rate* eine gute Leistung des Chirurgen bzw. der Institution in diesem Aspekt, so dass letztendlich eine patientenorientierte Information entsteht.

Ein weiterer Aspekt ist die Vermeidung unnötiger Operationen, so dass wir die *Negative Shunt Revision Rate* vorgeschlagen haben. Während eine gewisse Anzahl solcher Operationen im klinischen Alltag aufgrund diagnostischer Unsicherheit unvermeidbar ist, sollte aus unserer Sicht eine hohe Rate an negativen Shunt-Explorationen aufgrund von vermeintlichen hydrozephalen Beschwerden, die retrospektiv nach Operation zum Beispiel als Migräne-Kopfschmerz oder Erbrechen durch Gastroenteritis erkannt werden konnten, eine Verbesserung des klinischen Algorithmus mit sich bringen. Bei Vorliegen von klinischen Warnzeichen einer Shuntfehlfunktion oder bei diagnostischer Unsicherheit sollte im Zweifelsfall sicherlich weiterhin eine Exploration erfolgen, eine exzessiv hohe Rate an negativen Shuntrevisionen sollte aber zumindest eine Einzelfallanalyse und eine Re-Evaluation der Qualität der präoperativen Diagnostik bewirken. Bei Kindern mit unklaren Symptomen und zweifelhafter Diagnose einer Shuntfehlfunktion anhand klassischer klinischer und radiologischer Kriterien könnte die diagnostische Sicherheit vor einer operativen Shuntexploration beispielsweise durch ultrasonografische Messung des Optikusscheidendurchmessers oder durch invasive Hirndruckmessung erhöht werden.^{102–105}

In unserem Kollektiv betrug die *Negative Shunt Revision Rate* 7,1%. Wie bei anderen Indikatoren auch, muss der Referenzbereich letztendlich durch Reproduktion an weiteren Serien definiert werden.

Schlussfolgerung:

Die an unserem Patientenkollektiv bestimmten Qualitätsindikatoren für Shuntoperationen korrelieren mit den publizierten Referenzwerten und lassen sich mit vertretbarem Aufwand aus klinischen Routine-Daten bestimmen. Die

vorgeschlagene *Negative Shunt Revision Rate* deckt einen bisher nicht adressierten Aspekt ab und stellt eine patientenorientierte, sachlich sinnvolle, leicht erfassbare und potentiell modifizierbare Messgröße dar.

2.5) Headache and Shunt-related Impact on Activities of Daily Life in Patients Growing up with a Ventriculoperitoneal Shunt.

Beez T, Bellstädt L, Steiger HJ, Sarikaya-Seiwert S

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Einleitung:

Die Mehrzahl der als Kinder mit einem Liquorshunt versorgten Patienten verbleiben in Kindheit, Jugend und vermutlich auch Erwachsenenalter shuntabhängig.¹⁰⁶ Während der Einfluss von Shundysfunktionen und –infektionen gut untersucht worden ist, finden sich relative wenige Studien zu Lebensqualität und psychosozialem Einfluss bei Kindern, welche mit einem Shunt aufwachsen.^{63,107} Beispielsweise scheint auch bei suffizient therapiertem Hydrozephalus durchaus eine erhöhte Inzidenz von Kopfschmerzen bei Shuntpatienten im Vergleich zur Normalbevölkerung zu bestehen.¹⁰⁸ Je früher ein Shunt im Leben implantiert wird, desto größer kann der Einfluss solcher Faktoren auf die kognitive, soziale, akademische und berufliche Entwicklung sein, zusätzlich zu den Folgen einer Grunderkrankung. Ziel der vorliegenden Studie war es, die Belastung durch Kopfschmerzen sowie den Einfluss des Shunts auf Aktivitäten des täglichen Lebens an einem Patientenkollektiv im klinischen Alltag zu untersuchen, um praxisbezogene Erkenntnisse zu generieren.

Methoden:

Aus der Gruppe der an unserer Klinik zwischen 2005 und 2010 mit einem primären Liquorshunt versorgten Patienten jünger als 16 Lebensjahre wurden Kinder im Alter

von 3 (Kindergartenkinder) bis 21 Lebensjahren (Übergang in die neurochirurgische Erwachsenenversorgung) selektiert, welche zum Zeitpunkt dieser Studie wenigstens 1 Jahr Nachbeobachtungszeit seit der Operation aufwiesen. Die Patienten bzw. Familien wurden schriftlich und/oder telefonisch kontaktiert, ob sie an einer schriftlichen oder mündlichen Befragung teilnehmen würden.

Für die Befragung wurde einerseits ein nicht-validierter Fragebogen (Teil A) erstellt, welcher 7 Fragen zu Symptomkontrolle, zu Einfluss der Symptome und des Shunts auf den Alltag und auf spezielle Situationen (z.B. Urlaubsreisen) sowie zu technischen Problemen und Verbesserungsmöglichkeiten des Shunts aus Sicht der Betroffenen beinhaltet. Teil B bestand aus der deutschsprachigen Version des Strukturierten Kopfschmerzinterviews für Kinder (SIKI).¹⁰⁹ Dieses Interview richtet sich an das Kind und die Eltern und exploriert Symptome, Auslöser, *Coping*-Strategien, Emotionen und subjektives Krankheitsmodell in Bezug auf Kopfschmerzen. Die Auswertung erfolgte mittels deskriptiver Statistik.

Ergebnisse:

Vierzig Patienten wurden kontaktiert und 15 Patienten (9 männlich, 6 weiblich) stimmten der Befragung zu. Das mittlere Alter zum Zeitpunkt der Befragung lag bei 12 Jahren und die mittlere Nachbeobachtungszeit seit Shuntanlage bei 7 Jahren. Die Ätiologie des Hydrozephalus war kongenital (N = 8), tumorbedingt (N = 4) und posthämorrhagisch (N = 3). Das mittlere Alter bei Shuntimplantation lag bei 2 Jahren. In dem befragten Kollektiv kam es zu insgesamt 12 Shuntrevisionen bei 9 Patienten, wovon 10 aufgrund von Shuntokklusion und 2 aufgrund von Infektion notwendig waren.

In Teil A der Befragung gaben jeweils 87% der Befragten (N = 13) eine generelle Zufriedenheit mit dem Shunt und eine Verbesserung des Gesundheitszustandes durch den Shunt an. Die Mehrheit gab keine relevanten Einschränkungen im Alltag an (N = 10, 67%). Allerdings berichteten zwei Patienten von Schamgefühl bzw. erfahrenem Spott aufgrund des unter der Haut erkennbaren Shunts und fünf Patienten vermeiden Sport bzw. Anstoßen des Ventils. Die Hälfte der Befragten (N = 8, 53%) trifft besondere Vorkehrungen bezüglich Urlaubsreisen, wie zum Beispiel besonderes Augenmerk auf Flüssigkeitszufuhr und Sonnenschutz sowie Information über neurochirurgische Kliniken am Urlaubsort und Mitführen von Shuntausweis und medizinischen Dokumenten. Die Mehrheit (N = 12, 80%) forderte technische Verbesserungen des Shunts, besonders bezogen auf kleinere Ventile und verbesserte Zuverlässigkeit.

In Teil B der Befragung gab die Mehrheit Kopfschmerzen weniger als einmal pro Monat an (N = 10, 67%) und nur 3 Befragte (20%) leiden jede Woche unter Kopfschmerzen. Es handelt sich meist um Episoden mit einer Dauer unter 12 Stunden (N = 10, 67%). Der Beginn ist meist schleichend und es lässt sich keine tageszeitliche Zuordnung erkennen (N = 6, 40%). Typisch sind frontale Kopfschmerzen (N = 12, 80%) mit drückendem Charakter (N = 7, 47%) und einer mittleren Intensität auf der visuellen Analogskala von 5. Die Mehrheit muss niemals oder sehr selten aufgrund von Kopfschmerzen auf Schulbesuch oder Freizeitaktivitäten verzichten (N = 10, 67%). Die subjektive Einschätzung der Ursache der Kopfschmerzen war gleichmäßig verteilt: Shunt (N = 3, 20%), Dehydration (N = 3, 20%), Nackenverspannung (N = 2, 13%) und Wetterwechsel (N = 2, 13%). Die Frequenz der Kopfschmerzen war stabil oder besser werdend in 9 (60%) und die Intensität war stabil oder besser werdend in 10 (67%) Fällen.

Diskussion:

Die Hauptergebnisse dieser Studie sind die hohe generelle Zufriedenheit mit dem Shunt und subjektive Verbesserung des Gesundheitszustandes durch den Shunt in jeweils fast 90% der Fälle sowie die geringe Häufigkeit von alltagsrelevanten Kopfschmerzen.

Frühere Studien mit ähnlichen Kollektiven nutzten den *36-Item Short Form Health Survey* (SF-36) und Barthel Index oder den *Hydrocephalus Outcome Questionnaire* (HOQ) und *Health Utilities Index Mark 3* (HUI3).^{64,110,111} Diese Studien detektierten einen im Vergleich zur Normalbevölkerung reduzierten Gesundheitszustand bzw. eingeschränkte Lebensqualität bei Shuntpatienten. Als Determinante der Lebensqualität wurden u.a. Shuntstörungen identifiziert, also einen modifizierbaren Risikofaktor. Dies spiegelt sich in gewisser Weise auch in den Ergebnissen unserer Befragung wider, da viele der Befragten technische Verbesserungen, insbesondere auch der Zuverlässigkeit, forderten.

Kulkarni et al. verglichen die Lebensqualität nach Shuntanlage im Vergleich zur ETV und fanden keinen signifikanten Unterschied in der multivariaten Analyse.⁶⁸ Diese Ergebnisse könnten darauf hindeuten, dass die Ätiologie des Hydrozephalus bzw. die Grunderkrankung größere Bedeutung für die Lebensqualität hat als die Therapiemodalität.¹¹²

Kopfschmerzen waren in unserem Kollektiv kein bedeutsames alltagsrelevantes Symptom. Die Datenlage hierzu ist nicht eindeutig, da einige Autoren ebenfalls keine grundsätzlich erhöhte Kopfschmerzinzidenz bei Shuntpatienten erkennen konnten, andere Gruppen hingegen durchaus von einer gegenüber der Normalbevölkerung erhöhten Kopfschmerzhäufigkeit ausgehen.^{108,113} Auch unsere Ergebnisse legen nahe, dass vermutlich eine Subgruppe mit chronischen idiopathischen

Kopfschmerzen innerhalb des Shuntpatientenkollektivs besteht. Vor Diagnose einer chronischen idiopathischen Kopfschmerzerkrankung bei diesen Patienten muss allerdings aus unserer Sicht ein potentiell modifizierbares shunt-assoziiertes Problem, wie zum Beispiel Unterdrainage, Überdrainage oder Schlitzventrikelsyndrom, als Ursache sicher ausgeschlossen werden.^{113,114}

Schlussfolgerung:

Die Ergebnisse dieser Studie zeigen, dass moderne Shuntsysteme gut toleriert werden und durchaus bei der Mehrzahl der Kinder bzw. jungen Erwachsenen eine gute Lebensqualität erreicht werden kann. Die meisten Patienten passen ihren Lebenswandel an die Situation an und vermeiden subjektive Risiken und bereiten sich z.B. auf Urlaubsreisen gesondert vor. Chronische Kopfschmerzen stellen in der befragten Gruppe kein signifikantes Problem dar. Trotz grundsätzlicher Zufriedenheit besteht seitens der befragten Patienten aber technischer Verbesserungsbedarf, besonders in Bezug auf Größe und Zuverlässigkeit der Ventile bzw. Shunts.

3) Diskussion

3.1) Technische Aspekte bei der Implantation von Liquorshunts (Fragen und Ziele a bis d)

Unsere Studie konnte nachweisen, dass sich die Präzision der Platzierung des ventrikulären Katheters durch Nutzung des intraoperativen Ultraschalls verbessern lässt gegenüber einer Freihandpunktion.¹¹⁵ Wichtig erscheint die komplette Vermeidung von Fehllagen, welche mit dem höchsten Risiko einer späteren Shuntfehlfunktion einhergehen. Von den fehlplatzierten Kathetern in der Kontrollgruppe wurde bei 83% eine spätere Fehlfunktion mit entsprechender Revisionsoperation festgestellt. Insofern lässt sich durch verbesserte Lage des ventrikulären Katheters die Shunt dysfunktionsrate reduzieren. Vergleichbare Ergebnisse konnten auch mit der Neuronavigation erzielt werden.^{50,51,79} Zudem wurden kürzlich die Ergebnisse der Erprobung eines neuen intraoperativen Führungsinstruments (Thomale Guide, Christoph Miethke GmbH & Co. KG, Potsdam) mit Smartphone-gestützter Planungsapplikation mittels einer prospektiven randomisierten Studie publiziert, an welcher unsere Klinik als Studienzentrum beteiligt war.¹¹⁶ Die Planungsapplikation ermöglicht die Berechnung von patientenindividuellen Koordinaten, welche auf den Thomale Guide übertragen werden und somit intraoperativ eine präzise geführte Punktion des Ventrikels erlauben. Die Studie zeigte eine Verbesserung der Katheterlage und eine Verringerung der erforderlichen Punktionsversuche im Vergleich zur Freihandpunktion, allerdings keine Unterschiede in Bezug auf Katheterokklusionsrate oder Komplikationsrate.

Aus unserer Sicht sollte aufgrund dieser grundsätzlich positiven Ergebnisse eine technische Hilfe bei der Anlage von Ventrikelkathetern genutzt werden, was auch durch eine formale Meta-Analyse empfohlen wird.⁵³

Der Einfluss verschiedener Shuntventile auf das *Outcome* ließ sich in unserer Studie nicht nachweisen.¹¹⁷ In Abhängigkeit vom implantierten Shuntventil, insbesondere Differentialdruckventil versus Gravitationsventil, fanden sich im klinischen Alltag keine Unterschiede in der Überdrainage-Rate. Eine Einschränkung in der Verallgemeinerbarkeit unserer Ergebnisse ist das retrospektive Studiendesign mit Analyse eines institutionellen Algorithmus zur Ventilauswahl und entsprechendem Bias bei Alter und Ätiologie. Allerdings fand sich auch bei Nutzung eines randomisierten prospektiven Studiendesigns kein signifikanter Unterschied zwischen den getesteten Ventilen.⁴⁵ Eine evidenzbasierte Ventilauswahl erscheint somit nicht möglich, und man muss vermutlich auf theoretische oder finanzielle Aspekte sowie Erfahrungswerte oder Expertenmeinung zurückgreifen, was sich in der diesbezüglich stark variierenden Praxis erfahrener Kinderneurochirurgen widerspiegelt.¹¹⁸

Vielleicht zeigen zukünftige Ventilentwicklungen, z.B. elektromechanische *smart shunts* mit sensor-basierten Kontrollmechanismen, einen signifikanten Vorteil.¹¹⁹ Diese Systeme könnten die Liquorförderrate automatisch an den intrakraniellen Druck oder den Liquorfluss anpassen und Messdaten oder Warnungen vor sich anbahnenden Shuntproblemen z.B. kabellos an *Smartphones* übertragen.¹²⁰ Moderne Methoden werden zudem genutzt, um auch andere Komponenten eines Shunts zu optimieren. Galarza et al. nutzten *computational fluid dynamics* zur Entwicklung von Ventrikelkathetern mit optimiertem Flussprofil, welche eine niedrigere Okklusionsrate aufweisen sollen.¹²¹

3.2) Outcome der modernen Therapie des kindlichen Hydrozephalus mit Liquorshunts (Fragen und Ziele e bis h)

Mit Hilfe des von uns entwickelten Modells lässt sich Morbidität und Mortalität in der pädiatrischen Neurochirurgie mit moderatem Aufwand erfassen und anhand von Kenngrößen mit der Literatur vergleichen.¹²² Liquorassoziierte Eingriffe haben einen hohen Stellenwert, da sie einen großen Anteil kinderneurochirurgischer Operationen ausmachen und die Komplikationsrate dieser Eingriffe vergleichsweise hoch ist. Die Qualität speziell dieser Eingriffe lässt sich mit verschiedenen Qualitätsindikatoren messen, von denen einige aus unserer Sicht leicht erfassbar, sachlich sinnvoll, patientenorientiert und modifizierbar sind.¹²³ Allerdings wird vermutlich kein einzelner Indikator alle Aspekte abbilden können bzw. einige Indikatoren werden keinen klinischen Nutzen bringen und damit über die Bedeutung von „Gedankenspielen“ nicht hinauskommen.¹⁰¹ Welche Relevanz die von uns vorgeschlagene *Negative Shunt Revision Rate* in dieser Hinsicht haben wird, muss abgewartet werden.

Wir konnten zeigen, dass die Anwesenheit eines Liquorshunts bei heranwachsenden Menschen im Alltag in Bezug auf Einschränkungen sowie Symptomlast, insbesondere Kopfschmerzen, in der subjektiven Bewertung durch Betroffene keinen gravierenden Einfluss hat.¹²⁴ Allerdings passen die meisten Patienten ihren Lebenswandel an die Situation an und agieren teils vorsichtig bzw. vorbereitend. Die Befragten gaben technischen Verbesserungsbedarf besonders in Bezug auf die Zuverlässigkeit der Ventile bzw. Shunts an, was mit anderen Studien in Einklang steht, welche Shunt Dysfunktionen als fortwährende Quelle reduzierter Gesundheit bzw. Lebensqualität bis in das Erwachsenenalter identifizieren konnten.^{64,111,125}

4) Zusammenfassung

Der Hydrozephalus stellt eine Erkrankung mit zahlreichen Facetten dar, was Ätiologie, Manifestation, Therapie und *Outcome* in den verschiedenen Altersgruppen betrifft. Trotz der offenen Fragen, teilweise fundamentale Aspekte wie Physiologie der Liquorzirkulation oder Auswahl des Ventils betreffend, lässt sich festhalten, dass die Mehrzahl unserer Patienten mit dem aktuellen Stand der neurochirurgischen Hydrozephalus-Versorgung eine gute Lebensqualität erreichen kann. Dieser aktuelle Stand stützt sich auf zahlreiche wissenschaftliche Untersuchungen mit belastbaren Ergebnissen, was die Auswahl der optimalen Therapiemodalität, die Vermeidung von Infektionen und die Optimierung der intraoperativen Präzision angeht. Auf die Hydrozephalustherapie, speziell auf Liquorshunts, zugeschnittene Qualitätsindikatoren ermöglichen eine Sicherstellung und Optimierung der Patientenversorgung. Zukünftige theoretische und experimentelle Grundlagenwissenschaft, klinische Forschung und technische Innovationen werden die Therapie des Hydrozephalus noch weiter optimieren.

“Although we still have a long way to go in the practice of pediatric neurosurgery, we do more good for more patients by treating their hydrocephalus optimally than anything else.”

J. Gordon McComb¹

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Erklärung

Diese kumulative Habilitationsschrift vereint vom Verfasser publizierte Originalarbeiten, deren inneren wissenschaftlichen Zusammenhang der Themenkomplex des kindlichen Hydrozephalus darstellt. Die vorliegende Arbeit ist in Übereinstimmung mit der geltenden Habilitationsordnung der Medizinischen Fakultät der Heinrich-Heine-Universität Düsseldorf vom 02.09.2016 verfasst worden.

Ich versichere an Eides Statt durch meine Unterschrift, dass ich die vorliegende schriftliche Habilitationsleistung als eigenständige wissenschaftliche Leistung selbständig und ohne unzulässige fremde Hilfe angefertigt habe.

Bei den wissenschaftlichen Untersuchungen, die Gegenstand der schriftlichen Habilitationsleistung sind, wurden ethische Grundsätze und die Grundsätze und Empfehlungen zur Sicherung guter wissenschaftlicher Praxis beachtet.

Ich versichere, dass keine anderen eingeleiteten oder erfolglos beendeten Habilitationsverfahren bestehen.

Düsseldorf, den 24.10.2017

Dr. med. Thomas Beez

Real-time ultrasound guidance for ventricular catheter placement in pediatric cerebrospinal fluid shunts

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Abstract

Purpose Cerebrospinal fluid shunt failure is related to additional morbidity. Misplacement of ventricular catheters occurs in 40 % with freehand technique and is a risk factor for shunt failure. The goal of this study was to analyze the impact of intraoperative real-time ultrasound on catheter positioning and outcome in children.

Methods We compared children receiving ultrasound-guided procedures to matched historical freehand controls. Burr hole and convex probes were used (ProSound Alpha 6, Hitachi Aloka Medical Ltd., Tokyo, Japan). Catheter position was graded as grade I (optimal), II (contralateral ventricle or contact with ventricular structures), or III (misplacement). Correlation analysis was performed to identify determinants of outcome.

Results The study group ($n=17$) was balanced with the control group ($n=14$) for variables such as mean age (4.7 vs 4.3 years) and preoperative frontal occipital horn ratio (FOHR; 0.45 vs 0.43). In the study group, grade I catheter position was achieved in 6 (35 %) and grade II in 11 patients (65 %), compared to 2 (18 %) and 3 patients (27 %) in the control group. While no grade III position occurred in the study group, it was found in nine control patients (43 %) ($P=0.0029$). Failure rate was highest in grade III (83 %) compared to grade I catheters (50 %).

Conclusions This analysis demonstrated an improvement of catheter positioning with ultrasound guidance. In the absence of additional burden or risks, this method should be favored over freehand technique. It remains to be demonstrated in a

randomized controlled fashion to what extent improved catheter position translates into improved outcome.

Keywords Pediatric neurosurgery · Ventricular catheter · Hydrocephalus · Shunt failure · Intraoperative ultrasound

Introduction

Cerebrospinal fluid (CSF) shunts are a mainstay of therapy for hydrocephalus. In most patients, the shunt should be considered a lifelong implant due to persistent dependency on CSF diversion. Therefore, episodes of shunt failure represent a risk factor for additional morbidity and mortality, repeated surgical interventions and increased radiation exposure for the individual patient, and a financial burden for health services [23]. Several studies have for this reason examined determinants of shunt survival. While modifications of surgical technique were shown to achieve a reduction in infectious shunt failure [3, 18, 19], the influence of shunt hardware on failure rate remains debatable [2, 11]. Some studies suggest that optimal placement of the ventricular catheter might affect early shunt failure, which is predominantly caused by proximal malfunction [4, 10]. However, the most commonly used freehand technique for ventricular catheter placement is associated with a misplacement rate as high as 40 % [15]. Children are considered a special risk group due to the higher incidence of complex hydrocephalus, small ventricles, or aberrant ventricular anatomy. In this study, we have thus analyzed the impact of real-time ultrasound guidance on the precision of ventricular catheter placement and on outcome in a pediatric cohort.

TB and SSS contributed equally to the study.

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Materials and methods

Patients

After obtaining approval by the local institutional review board (study no. 4588), we identified all patients younger than 18 years on whom ultrasound-guided placement of a ventricular catheter was performed at our institution between April 2012 and March 2013. The control group was matched on the basis of age from the patient cohort treated before introduction of intraoperative ultrasound at our institution, when ventricular catheters were exclusively placed by freehand technique. Ventricular size and catheter position were assessed on routine cranial imaging (computed tomography or magnetic resonance imaging). Ventricular size was quantified by calculating the frontal occipital horn ratio (FOHR) as previously described [16]. Ventricular catheter position was determined on first postoperative imaging using the following grading system, which was modified from a previous report [5]: Grade I corresponds to an optimal position with the catheter tip being surrounded by CSF in the ipsilateral ventricle. Grade II summarizes all other intraventricular positions, i.e., the catheter tip located in the contralateral ventricle or touching ventricular wall, septum pellucidum, or choroid plexus. Grade III refers to misplaced catheters, defined as catheter tip or whole catheter lying outside of the ventricular system.

Surgical technique

In children without patent fontanelles, a cranial burr hole was made using a 14-mm disposable perforator (Codman & Shurtleff Inc., Raynham, USA). Prior to opening of the dura, the burr hole was extended laterally by 3–5 mm with a

Kerrison rongeur, as depicted in Fig. 1. The dura was opened within the lateral extension using a bipolar forceps. An ultrasound console (ProSound Alpha 6, Hitachi Aloka Medical Ltd., Tokyo, Japan) with a burr hole probe 3.0–7.5 MHz (UST-52114P, Hitachi Aloka Medical Ltd., Tokyo, Japan) was used. The burr hole probe was inserted into the burr hole with the probe's guide rail directed toward the lateral extension (Fig. 1). After adjusting the probe to the desired trajectory, the neurosurgeon inserted the ventricular catheter through the probe's guide rail. A representative ultrasound view obtained with the burr hole probe is presented in Fig. 2.

In children with patent fontanelles, a convex probe 3.75–10 MHz (UST-9120, Hitachi Aloka Medical Ltd., Tokyo, Japan) was used for continuous real-time ultrasound visualization of the procedure via the fontanelle. A typical ultrasound view obtained via the fontanelle is presented in Fig. 2.

The unique feature of ultrasound-guided placement of ventricular catheters is the continuous real-time visualization of the catheter advancing through the parenchyma into the desired ventricular compartment, allowing for active correction of the catheter's trajectory during puncture. The surgical setting as well as the dynamic nature of this procedure is represented in Fig. 3.

Statistical analysis

Descriptive statistics were used for quantitative analysis of data. Values are presented as means unless specified otherwise. Fisher's exact test, with Freeman-Halton extension where applicable, was performed to statistically examine association between groups and outcomes. *P* values are two-tailed and the level of significance was determined a priori to be $P < 0.05$. All analyses were performed by using Prism and

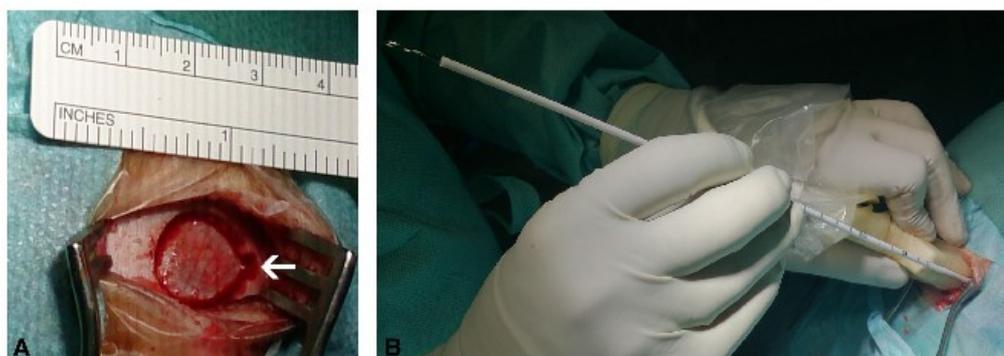
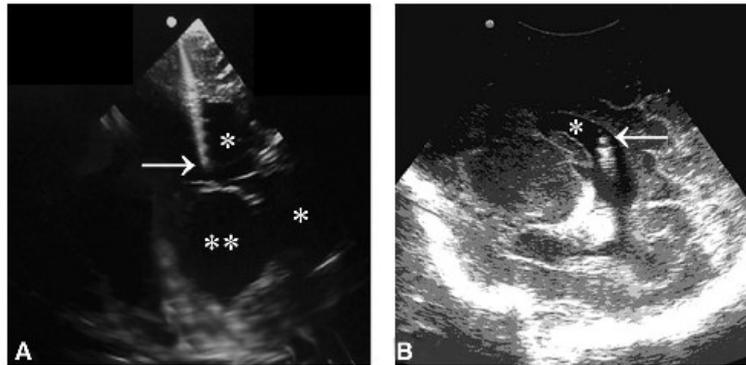


Fig. 1 **a** In children without patent fontanelles, a cranial burr hole was made using a 14-mm disposable perforator (Codman & Shurtleff Inc., Raynham, USA). Arrow indicates a lateral extension by 2–3 mm made with a Kerrison rongeur, to allow for the ventricular catheter passing beside the burr hole probe. **b** Handling of probe and catheter for real-time

ultrasound in children without patent fontanelles: The burr hole probe was inserted into the burr hole with the probe's guide rail directed toward the lateral extension, where the ventricular catheter is inserted through a small dural incision

Fig. 2 **a** Representative ultrasound view after ventricular catheter insertion obtained with the burr hole probe in a child without patent fontanelle. *Single asterisks* indicate the lateral ventricles; *double asterisk* indicates the third ventricle. **b** Representative ultrasound view after ventricular catheter insertion obtained with the convex probe in a child with patent fontanelle. *Asterisk* indicates the lateral ventricle; *arrow* indicates the tip of the ventricular catheter



QuickCales statistical software (GraphPad Software, San Diego, USA).

Results

Patient and shunt data

Seventeen children underwent ultrasound-guided placement of a ventricular catheter for CSF shunts. The mean age in this study group was 4.7 years (range 0 to 15 years). The control group comprised 14 patients with a mean age of 4.3 years (range 0 to 17 years). Both groups were well balanced with regard to underlying pathology and implanted shunt hardware,

as presented in Table 1. Mean preoperative FOHR was 0.45 (range 0.34 to 0.71) in the study group and 0.43 (range 0.29 to 0.66) in the control group. Mean duration of surgery was 51 and 49 min, respectively. The mean follow-up was 10.4 months (range 3 to 16 months) in the study group and 27.6 months (range 1 to 84 months) in the control group.

Accuracy of ventricular catheter placement

In the study group, a grade I catheter position was achieved in 6 patients (35 %) and a grade II position in 11 patients (65 %), as presented in Table 2. There was no case of a grade III catheter position. In the control group, two patients (18 %) showed a grade I position, three patients (27 %) a grade II

Fig. 3 Artist's rendition giving an overview of real-time ultrasound-guided ventricular catheter placement (*upper right*). *Panel A* depicts the burr hole probe, catheter, and burr hole. *Panel B* depicts convex probe, catheter, and fontanelle. *Panels 1–3* illustrate the procedure in a step-wise fashion from left to right: Ultrasound pictures on the *left* show the catheter advancing through the brain parenchyma into the ventricle. The corresponding illustration of the surgical handling is represented in the *right pictures*, respectively

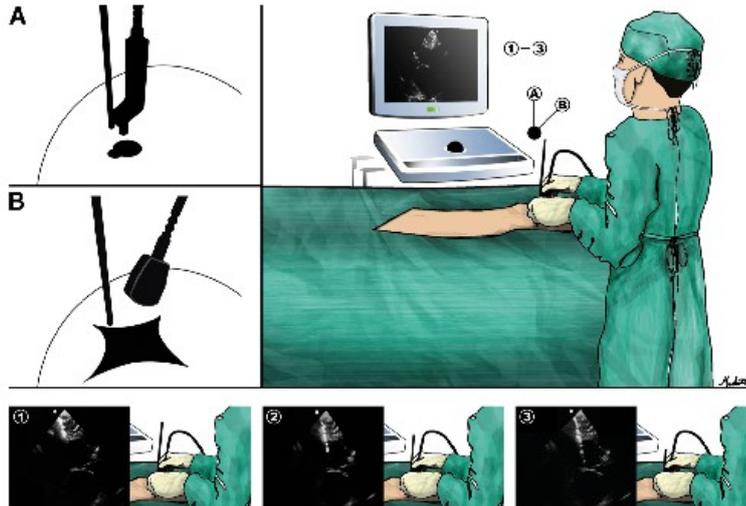


Table 1 Patient and shunt data

	Study group (ultrasound guidance)	Control group (freehand technique)
No. of patients	17	14
Mean age (range)	4.7 years (0–15 years)	4.3 years (0–17 years)
Etiology of hydrocephalus:		
Posthemorrhagic	10 (59 %)	5 (36 %)
Congenital	3 (18 %)	3 (21 %)
Dysraphic	2 (12 %)	1 (7 %)
Tumor	1 (6 %)	2 (14 %)
Other	1 (5 %)	3 (21 %)
Valve types:		
paediGAV (gravitational)	12 (71 %)	8 (57 %)
Medos Hakim (adjustable)	1 (6 %)	2 (14 %)
Codman Certas (adjustable)	4 (24 %)	0 (0 %)
Other	0 (0 %)	4 (29 %)

Groups were matched for age and are well balanced regarding etiology of hydrocephalus and valve type

position, and six patients (55 %) a grade III position (Fig. 4). The association between groups and outcomes, i.e., catheter position grading, was analyzed using the Freeman-Halton test with the null hypothesis being that ultrasound intervention does not affect catheter position. With $P=0.0029$, the null hypothesis was rejected, concluding that ultrasound intervention had a statistically significant effect on catheter position grading. In three patients in the control group, catheter position could not be reliably assessed due to a lack in adequate postoperative imaging. There was no statistically significant difference between ultrasound guidance via burr hole versus patent fontanelle (data not shown).

Influence on shunt failure rates

Five patients (29 %) within the study group experienced shunt failure. Three failures were attributed to proximal dysfunction and two failures to infection. The mean shunt survival was

Table 2 Accuracy of ventricular catheter placement

	Study group (ultrasound guidance)	Control group (freehand technique)
Grade I	6	2
Grade II	11	3
Grade III	0	6

Freeman-Halton exact test $P=0.0029$

Statistical analyses revealed a significant difference regarding catheter positions, with more precise positions being achieved in the study group

10.4 months (range 3 to 16 months). In the control group, shunts failed in nine patients (64 %). Five of these patients (35 %) experienced shunt failure within the first year after implantation (Fig. 5). The mean shunt survival was 27.6 months (range 1 to 84 months). In eight patients, the shunt failed due to proximal dysfunction, whereas in one case, infection occurred. When analyzing shunt failures due to proximal dysfunction, we found a tendency toward highest failure rate (83 %) in grade III catheters ($n=5$). Detailed results are presented in Table 3. Differences did not reach statistical significance.

Discussion

Ultrasound-guided application of ventricular catheters significantly improved the precision of placement in this study and completely eliminated misplacement outside of the ventricular system, i.e., grade III catheter positions. In the ultrasound group, 100 % of catheter tips were found to be in a grade I or II position, compared to only 45 % in the freehand group. Analysis of shunt survival rates did not reveal a significant advantage of ultrasound-guided placement with regard to clinical outcome. Of note, the proportion of shunt failure tended to be higher in grade III catheters.

Although the freehand technique is thought to be most commonly applied method of ventricular catheter insertion, its considerable rate of inaccurate punctures is well documented. Lind et al. compared the accuracy of different approaches for ventricular catheter insertion by freehand technique [15]. They found that only 64 % of frontal and 42 % of occipital catheters were placed within the predefined ventricular target zones. In a series of 129 external ventricular drains placed by means of freehand technique, 64.3 % of catheters were placed correctly within the ipsilateral ventricle [8]. The most common suboptimal position was the contralateral ventricular system, whereas 14 % of catheters were completely misplaced in the brain parenchyma. Unfortunately, the authors did not present a detailed analysis of catheter position, which hampers comparison of data sets. They additionally reviewed 10 published studies on freehand ventricular catheter placement and confirmed a mean rate of inaccurate catheter positions, i.e., outside of the ventricular system, in approximately 15 % of cases.

In contrast, most studies investigating technical assistance to improve ventricular catheter positions report enhanced precision and reduction of cases with extraventricular placement. Several authors analyzed the impact of electromagnetic-guided navigation [1, 5, 6, 9, 14, 27] as well as stereotactic guidance with rigid head fixation [20] and came to positive results. Neuroendoscopy was also investigated in this context, although findings are somewhat divergent [12, 22]. Different

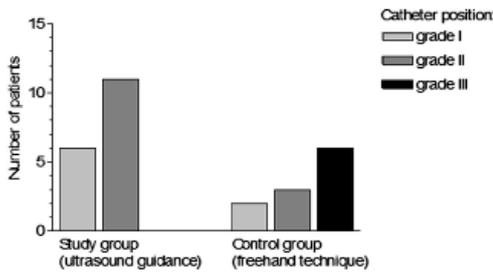


Fig. 4 Association between groups and outcomes was found to be statistically significant ($P=0.0029$), with no misplacements (i.e., grade III) occurring in the study group

types of guiding instruments have been evaluated and a smartphone-assisted guide is subject to an ongoing prospective trial [17, 21]. Regarding ultrasound guidance, technical reports on using a burr hole probe [24] as well as a conventional probe via the fontanelle [7] have been published. However, data on larger ultrasound cohorts is conflicting with one retrospective series demonstrating an improvement in precision [27], whereas a recent prospective trial failed to establish a significant clinical benefit of ultrasound guidance probably attributed to catheter movement in the postoperative course [25, 26]. With regard to clinical significance of such findings, the relation between morphologically correct catheter position and shunt survival has to be analyzed. In the present study, the shunt failure rate in the ultrasound group was 29 %, which is in accordance to the literature [13, 26]. The overall failure rate in the control group was higher, but a significant difference in follow-up time has to be taken into

account due to the retrospective methodology. When only looking at the failure rates at 1 year after surgery, when most proximal shunt failures are known to occur [10], we found a rate of 29 % in the study group compared to 35 % in the control group. Thus, we did not reveal a statistically significant difference between the ultrasound and freehand groups with regard to shunt failure rates. When reviewing the relevant literature, several studies suggest a reduction in early shunt failure due to proximal dysfunction when enhancing the precision of catheter placement by means of technical assistance [6, 9, 20, 27]. Of note, a prospective study on this topic demonstrated a significant difference in catheter position and also in failure rate when comparing optimal and poor catheter positions [5], whereas two prospective randomized trials of endoscopy and ultrasound failed to prove a benefit in outcome [12, 26]. These equivocal findings could be related to differences in study designs and outcome parameters, but of course also to varying abilities of the devices and procedures being tested.

In summary, real-time ultrasound significantly improved the precision of ventricular catheter placements in this series. The method allows for real-time visualization of the ventricular anatomy and of the catheter passing through the brain parenchyma into the ventricle. The additional burden for the patient is negligible, and neither additional preoperative imaging nor head fixation are required. The duration of surgery is not significantly longer, although preoperative preparation time for sterile draping of the ultrasound machine and probe has to be taken into account. However, the relation between catheter position and shunt failure rate remains unclear based on our data. Some limitations of our study are related to its

Fig. 5 Kaplan-Meier curves showing no differences in shunt survival rates between ultrasound and control groups in the first year after implantation

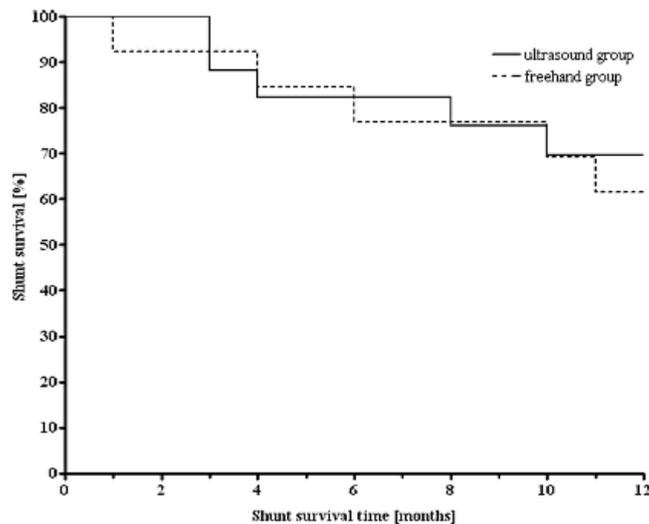


Table 3 Results of follow-up

	Study group (ultrasound guidance)	Control group (freehand technique)
Mean follow-up	10.4 months (3–16 months)	27.6 months (1–84 months)
Revisions	5 (29 %)	9 (64 %) [5 (35 %) in the first year]
Reason for shunt failure:		
Dysfunction	3 (60 %)	8 (89 %)
Infection	2 (40 %)	1 (11 %)
Failure due to dysfunction related to catheter grading:		
Grade I	3 (50 %)	1 (50 %)
Grade II	0 (0 %)	1 (33 %)
Grade III	0 (0 %)	5 (83 %)

Revision rates were similar between both groups in the first year and are generally in keeping with the literature. Failure rate was highest in grade III positions

retrospective design. There is a significant difference in follow-up time between the study and control groups. This bias can be avoided when only analyzing early shunt failure, which represent the majority of proximal dysfunctions [10]. Moreover, we were not able to reproduce the number of passes required for successfully placing the ventricular catheter due to the retrospective study design and unreliable operative notes regarding this particular aspect. In our experience, the learning curve for this method is steep, and an experienced neurosurgeon requires only one attempt in the majority of patients. Interestingly, as previously mentioned, the ultrasound technique performed in the study of Whitehead et al. was unable to improve catheter position, which the authors attribute to secondary catheter movement between intraoperative and postoperative imaging [25, 26]. It was unfortunately not possible to reassess this aspect in our cohort, although this phenomenon certainly deserves special attention in future trials on auxiliary means for improvement of ventricular catheter placement accuracy.

Real-time ultrasound guidance improved the accuracy of ventricular catheter placements for CSF shunts in this pediatric series and completely eliminated severe misplacements. Although risk of failure tended to be higher in poorly placed catheters, it remains to be assessed in a randomized controlled fashion to what extent improved catheter position finally translates into improved outcome for the patient. We conclude that ultrasound potentially offers clinically relevant advantages without additional burden for the patient and should thus be favored over freehand technique for placement of ventricular catheters.

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Conflict of interest The authors declare no financial or material support for conducting this study.

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Role of ventriculoperitoneal shunt valve design in the treatment of pediatric hydrocephalus – a single center study of valve performance in the clinical setting

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Abstract

Purpose Previous studies have established risk factors for ventriculoperitoneal shunt failure in children. However, the role of valve type as a determinant of complications and outcome remains unclear. The aim of this study was to compare the fixed-pressure paedigAV and the programmable Codman Hakim valves in the clinical setting.

Methods We conducted a retrospective review of patients younger than 16 years who underwent primary implantation of a ventriculoperitoneal shunt with either valve type at our institution between January 2005 and December 2010. Shunt survival analyses were performed to identify variables associated with risk of shunt failure.

Results Of the 44 patients in the paedigAV cohort, 50 % reached the endpoint of shunt failure with a mean time to shunt failure of 7 months. The Codman Hakim cohort comprised 29 patients, of which 55 % experienced shunt failure with a mean time to shunt failure of 8 months. Stratified analyses identified young age at implantation and posthemorrhagic hydrocephalus as risk factors for shunt failure. Shunt survival analysis revealed no significant difference with regard to valve type.

Conclusions This study confirmed important risk factors for shunt failure in children. Despite certain limitations and biases, similar findings for both valves examined in the clinical setting were obtained. Thus, valve type does not seem to influence risk of shunt failure. Prospective, randomized, and controlled trials are required to validate these results.

Keywords Hydrocephalus · Ventriculoperitoneal shunt · Children · Valve type · Complications · Shunt failure

Introduction

Pediatric hydrocephalus is considered a surgical disease, as the prevailing treatment is the implantation of a ventricular shunt [21]. The disease is multifaceted in many aspects: etiology of pediatric hydrocephalus is diverse, age groups are distinctly different concerning surgical challenges, risks and outcome, and finally management protocols and available shunt hardware are manifold [1]. This is also reflected by the existing literature, which consists of many retrospective and few prospective studies, as recently summarized [21]. Outcome variables and definitions vary widely and this heterogeneity often hampers comparison of results. However, certain correlations are well established: Young age at surgery, prematurity, posthemorrhagic, and postinfectious hydrocephalus are associated with an increased risk for shunt failure and infection, with a higher incidence of neurological deficits and cognitive impairment as well as with a lower degree of social integration and quality of life [21]. Concerning surgical technique, correct handling, and implantation technique of shunt hardware have been shown to influence shunt failure and infection rates [5, 11, 12]. Another relevant field is shunt technology, as this might be another major determinant of proper shunt function. However, even the largest trial comparing failure rates of different shunt valves revealed no significant differences between valve designs and thus provides no basis for evidence based decisions in this matter [4]. At our institution, pediatric shunt implantations are almost exclusively performed by a team of neurosurgeons experienced in the treatment of children. The standard procedure for ventriculoperitoneal shunting in children comprises little or no shaving, repeated antiseptic scrub of the whole operative

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field, curvilinear scalp incision, and straight abdominal incision without intermediary incisions, no-touch technique for shunt hardware, and ultrasound-guided placement of the ventricular catheter. With regard to shunt hardware, two types of valve are used: The fixed-pressure gravity-assisted Miethke paedigAV 4/19 and 9/19 valves (Aesculap AG & Co. KG, Tuttlingen, Germany) are mainly implanted in preterm and younger children due to the valve's small dimensions and propagated reduction in overdrainage-related problems. The programmable differential pressure Codman Hakim valve (Codman & Shurtleff, Inc., Raynham, USA) is preferentially chosen for older children due to its larger size and possibility of noninvasive adjustment of pressure settings. As this protocol is, at least concerning choice of valve type, rather based on experience than on scientific evidence, we conducted this retrospective study to compare the paedigAV and Codman Hakim valves in the clinical setting.

Methods

Patient population

We retrospectively identified all neurosurgical patients younger than 16 years who underwent primary implantation of a ventriculoperitoneal shunt at our institution between January 2005 and December 2010 and for whom either follow-up on a regular basis was at least 24 months or an endpoint of first shunt failure was recorded. From this cohort, patients who received either a Miethke paedigAV 4/19 or 9/19 valve or a programmable Codman Hakim valve were selected. Patient records were obtained from the institution's computerized database and anonymized prior to scientific analysis. The following data was extracted: etiology of hydrocephalus, age at implantation, type of valve, time of first shunt failure, and reason for shunt failure. Etiology was categorized into tumor (occlusive hydrocephalus, persisting hydrocephalus after tumor resection), hemorrhage (posthemorrhagic hydrocephalus after intraventricular hemorrhage and hydrocephalus after SAH), malformation (dysraphic and nondysraphic), and others (posttraumatic, postinfectious, and unspecified hydrocephalus). Shunt failure was defined as a malfunction making reoperation necessary. We distinguished between mechanical shunt failure (obstruction, disruption, and dislocation), functional shunt failure (overdrainage), as well as septic shunt failure. The latter was defined as a medical diagnosis based on unambiguous clinical findings with or without positive microbiological cultures, which leads to reoperation [21].

Statistical analyses

Descriptive statistics were used for quantitative analysis of the data. Nonparametric test statistics, including Mann-Whitney

and Kruskal-Wallis tests, were used to compare two or more groups, respectively. The Kaplan-Meier method was applied to analyze shunt survival with respect to age, etiology, and valve type. The level of significance for all statistical tests was determined to be $p < 0.05$ (two-tailed).

Results

Patient population

One hundred two patients younger than 16 years who underwent primary implantation of a ventriculoperitoneal shunt at our institution between January 2005 and December 2010 were identified. Of these patients, 44 (43 %) received a paedigAV valve and 29 (28 %) received a Codman Hakim valve. Thus, 73 patients were included in further analyses. Of these patients, 39 (53 %) reached the endpoint of shunt failure and revision surgery. Mean overall follow-up was 34 months (0–89 months). One patient died of neonatal sepsis and three patients died of brain tumors. There were no shunt-related deaths in the paedigAV and Codman Hakim cohorts.

Cohorts by valve

In the paedigAV cohort, 24 patients received a valve with opening pressure combination 4cmH₂O/19cmH₂O and 20 patients with opening pressure combination 9cmH₂O/19cmH₂O. The mean age on implantation in this group was 18 months (0–154 months), with 33 patients being ≤ 12 months of age, 5 patients being between 13 and 60 months, and 6 patients being > 60 months. Indications for shunt implantation were tumor in 11 (25 %), hemorrhage in 13 (39 %), malformation in 16 (36 %), and others including posttraumatic and postinfectious hydrocephalus in 4 patients (9 %), respectively. Twenty-two patients (50 %) reached the endpoint of shunt failure. The mean time until shunt failure was 7 months (0–56 months). Reason for shunt failure was mechanical in 14 (64 %), infectious in 6 (27 %), and functional in 2 patients (9 %), respectively. Of note, both cases of functional shunt failure were related to overdrainage with chronic subdural hematoma. One patient in this cohort died from tumor progression, and a further patient died from sepsis.

In the Codman Hakim cohort, the mean age on implantation was 70 months (1–180 months), with 4 patients being ≤ 12 months of age, 11 patients being between 13 and 60 months, and 13 patients being > 60 months. Indications for shunt implantation were tumor in 13 (45 %), hemorrhage in 4 (14 %), malformation in 8 (28 %), and others including posttraumatic and postinfectious hydrocephalus in 4 patients (13 %), respectively. Sixteen patients (55 %) reached the endpoint of shunt failure. The mean time until shunt failure was 8 months (0–42 months). Reason for shunt failure was

mechanical in 10 (59 %), infectious in 4 (24 %), and functional in 2 patients (12 %), respectively. Again, overdrainage was responsible for both cases of functional shunt failure. In this cohort, two patients died from tumor progression. Data from both cohorts is summarized in Table 1.

Stratification by age group

The etiological subgroup others was not included in this analysis, as no statistically meaningful assumptions could be made due to the very small patient number. When stratifying patients with the prevailing indications, i.e., tumor, hemorrhage, and malformation; for age, 36 patients underwent their primary operation at an age ≤12 months, 14 between 13 and 60 months and 15 at an age >60 months. In these age groups, 23 (64 %), 6 (43 %), and 6 patients (40 %) reached the endpoint of shunt failure. This trend towards higher revision rate of shunts implanted in very young children did not reach statistical significance. The corresponding shunt survival curves generated with the Kaplan Meier method are presented in Fig. 1.

Stratification by indication for primary shunt implantation

Twenty-five patients received their shunt for tumor-related hydrocephalus, 17 for posthemorrhagic hydrocephalus, and 24 for hydrocephalus associated with malformation. The endpoint of shunt failure was reached in 8 (32 %), 15 (88 %), and 12 patients (50 %), respectively. Kruskal Wallis test followed by Dunn's multiple comparison test showed a significantly higher shunt failure rate in patients with hemorrhage as

Table 1 Characteristics of paediGAV and Codman Hakim valve cohorts.

	paediGAV valve	Codman Hakim valve
No. of patients	44	29
Mean age on implantation	18 months (0-154)	70 months (1-180)
Etiology of hydrocephalus (%):		
Tumor	11 (25)	13 (45)
Hemorrhage	13 (30)	4 (14)
Malformation	16 (36)	8 (28)
Other	4 (9)	4 (14)
No. of patients with shunt failure	22 (50)	16 (55)
Mean time to shunt failure	7 months (0-56)	8 months (0-42)
Reason for shunt failure (%):		
Mechanical	14 (64)	10 (59)
Infectious	6 (27)	4 (24)
Functional	2 (9)	2 (12)

Age on implantation	No. of patients	No. of pat. with shunt failure	Shunt failure rate
≤12 months	36	23	64%
13-60 months	14	6	43%
>60 months	15	6	40%

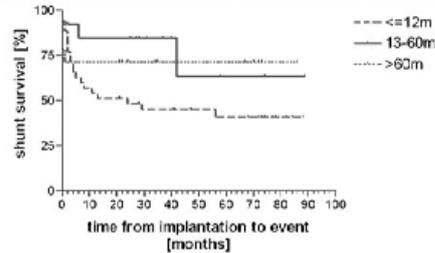


Fig. 1 Stratification by age group

underlying etiology ($p=0.0017$). The corresponding shunt survival curves are presented in Fig. 2.

Stratification by valve type

When stratifying patients with the prevailing indications, i.e., tumor, hemorrhage, and malformation for valve type, 40 patients received a paediGAV and 25 Codman Hakim valve. In the paediGAV group, 21 shunt failures (53 %) occurred compared to 14 (56 %) in the Codman Hakim group. Shunt survival curves are presented in Fig. 3.

Discussion

In this study, we compiled a retrospective single center series of children receiving ventriculoperitoneal shunts. We were able to confirm two established major risk factors for shunt failure, which are young age at implantation and hemorrhage

Indication for implantation	No. of patients	No. of pat. with shunt failure	Shunt failure rate
tumor	25	8	32%
hemorrhage	17	15	88%*
malformation	24	12	50%

* $p = 0.0017$

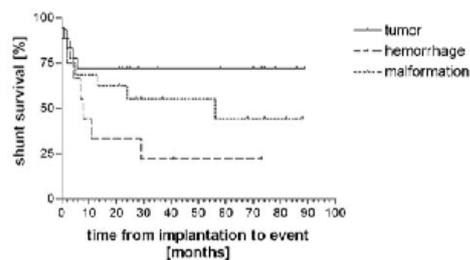


Fig. 2 Stratification by indication for primary shunt implantation

Valve type	No. of patients	No. of pat. with shunt failure	Shunt failure rate
paediGAV	40	21	53%
Codman Hakim	25	14	56%

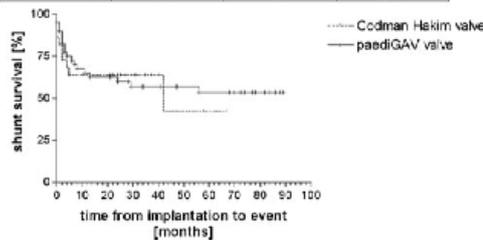


Fig. 3 Stratification by valve type

as underlying etiology. The two factors might be interrelated, as posthemorrhagic hydrocephalus mainly occurs in premature children. However, similar results were found in many previous studies [16, 20, 21]. Our shunt survival analysis also shows that the majority of reoperation for shunt failure occurs within the first year after implantation. This finding corresponds to a proportion of 75 % of reoperations performed within 1 year in a previous study of paediGAV in hydrocephalic children [7]. The shunt survival rates calculated in our study are similar to the results obtained in a recent analysis of children receiving the adjustable proGAV valve, which found an overall rate of 64.3 % and a rate of 60.9 % for infants aged <1 year [19].

The major aim of this study was to compare the paediGAV and Codman Hakim valves in the clinical setting. When reviewing these two cohorts, the distinctly different age on implantation becomes evident. This difference, which is highly relevant for interpreting our results, is clearly related to our institutional protocol. paediGAV valves are preferentially implanted in premature and younger children due to the valve's small dimensions, whereas the larger Codman Hakim valve is considered more adequate for older children. The different age distribution biases the results of this comparison, as younger age is by itself associated with increased risk for shunt failure. Additionally, this difference implies an imbalance in etiology of hydrocephalus with more posthemorrhagic hydrocephalus cases in the paediGAV cohort and more tumor cases in the Medos Hakim cohort. However, when stratifying for valve type regardless of age and etiology, we found identical revision rates for both valves. The mean shunt survival time as well as the reasons for shunt failure was almost identical. From a pragmatic point of view, our findings might be interpreted as similar performance of paediGAV and Codman Hakim valves in this specified clinical setting, of course taking into account the potential bias resulting from the acknowledged differences between the cohorts. Therefore, neither our work nor previous randomized controlled as well as retrospective studies succeeded in demonstrating superiority of one

particular shunt valve concerning shunt failure [4, 18]. The latter study even looked at long-term follow-up and found no significant influence of valve type on revisions occurring more than 10 years after implantation.

Interestingly, the rate of overdrainage in our series was similar between paediGAV and Codman Hakim cohorts and accounted for 9 and 12 %, respectively. Laboratory tests of various valve constructions published in 1995 suggested that the compensation of postural changes is a major technological challenge, which could not be met at that time [2]. In a large series of 169 children who received a paediGAV valve, overdrainage occurred in eight cases or 4.7 % [7]. The first published larger series of children treated with Codman Hakim valves comprised 78 patients and reported overdrainage in eight children or 10 % [14]. These results were subsequently confirmed by a randomized controlled study [13]. For other valve types, similar results were obtained, particularly in the randomized controlled shunt design trial, which compared two types of complex valves with ordinary differential pressure valves and found no statistically significant differences [4, 8]. In a series of 144 patients receiving a proGAV valve, which is an adjustable differential pressure valve combined with a gravitational unit, 35 cases of overdrainage were reported [17]. Adjustment of the valve was an effective treatment in all but one patient, who required ligation of the shunt. However, the opening pressure of the proGAV was decreased in 66 % and increased in 34 % of all adjustments performed within the study period. This finding implies that the main reason for adjustment was underdrainage rather than overdrainage. In this context, the authors themselves address an issue which is in our opinion highly important when reporting data on over- or underdrainage. In their multicenter study, there were no defined operational terms for drainage status and the decision to change pressure level was left at the discretion of each investigator. Overdrainage can be diagnosed radiologically and clinically. Extra-axial fluid collections [3], slit-like ventricles [15], and reproducible orthostatic headache [6] are reported manifestations of overdrainage. In our retrospective study, we were unable to reliably identify patients with overdrainage treated nonsurgically and referring to the Codman Hakim cohort cases in which pressure levels were adjusted for this reason. We are therefore only able to report the rate of severe symptomatic overdrainage causing extra-axial fluid collections and requiring revision surgery. This operational definition of overdrainage justifying surgery has been used before [9, 10].

Our study has the typical limitations of a retrospective study design. We only examined implantation and first shunt failure so that no data regarding repeated shunt failures are reported. Bias might arise from our institutional treatment protocol, which strongly influenced the characteristics of the cohorts available for analysis. However, we succeeded in compiling a pediatric hydrocephalic patient series giving

valuable information about the performance of paediatric GAV and Codman Hakim valves in every day clinical practice. With the study's limitations and biases in mind, we nevertheless achieved the aim of a clinically meaningful comparison between these two valve types. Moreover, several relevant findings from previous studies are confirmed.

Conclusions

Our study confirms the increased risk of shunt failure associated with young age at implantation and hemorrhage as underlying etiology of hydrocephalus. The risk for shunt failure is highest within the first year after implantation. Mechanical shunt failure, which includes obstruction, disruption, and dislocation, is the major reason for reoperation. Risk of shunt failure and especially incidence of overdrainage do not appear to be influenced by valve type. The similarity of findings for both examined valves in the clinical setting might support our institutional protocol for valve choice, which is primarily based on patient age, and does not indicate a major role of valve design with respect to the failure rate, of course with potential biases in mind. However, due to the limitations of this retrospective study, these findings should be further assessed in prospective, randomized, and controlled trials.

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Prospective review of 30-day morbidity and mortality in a paediatric neurosurgical unit

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Abstract

Purpose The purpose of this study is to record the 30-day and inpatient morbidity and mortality in paediatric patients in a tertiary neuroscience centre over a 2-year period. The intentions were to establish the frequency of significant adverse events, review the current published rates of morbidity in paediatric neurosurgical patients and propose three clinical indicators for future comparison.

Methods All deaths and adverse events were prospectively recorded from 1 January 2014 to 31 December 2015. Each adverse event was categorised, allocated a clinical impact severity score and linked to a neurosurgical procedure wherever possible. Where a patient suffered several adverse events in the same admission, each event was recorded separately. If a patient had been discharged home, an adverse event was recorded if it occurred within 30 days of admission.

Results Five hundred forty-nine procedures were performed in 287 patients (aged <16 years). One hundred thirty significant adverse events were identified. The following are the three clinical indicators: significant adverse event rate: 111 (20.2%) operations were linked to at least one significant adverse event; unscheduled return to theatre rate: 81 (14.8%) operations were associated with an adverse event that resulted in an unscheduled return to theatre; and surgical site infection rate: 29 (5.3%) operations were associated with an infection.

Conclusion Complications and adverse events are common in paediatric neurosurgery. Prospective, continuous surveillance will promote both quality assurance and quality improvement in the neurosurgical care delivered to patients.

Keywords Adverse event · Complication · Paediatric · Neurosurgery · Surveillance

Introduction

The French vascular surgeon Rene Leriche [7] famously wrote:

“Every surgeon carries about him a little cemetery, in which from time to time he goes to pray, a cemetery of bitterness and regret, of which they seek the reason for certain of their failures”.

In recent years, there has been much greater scrutiny of adverse events in healthcare systems and a surgeon’s personal cemetery is no longer private but to be opened for public scrutiny.

Individual consultant surgeon- and institution-specific mortality rates have been published in the UK since 2014. When first published, the NHS Medical Director, Sir Bruce Keogh, said that surgeons had a moral responsibility to make public their death rates, in arguing that this was a means to defend how well they delivered a service; he directly linked surgical outcome data to quality and safety.

Morbidity and mortality should be recorded in a systematic way; in addition to recording any adverse event, the system must also define the denominator—how often could such an event have occurred, to determine the true rate. When comparisons can be made between units, the standard of care can be defined.

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We sought to devise a system for the prospective surveillance of all morbidity and mortality in paediatric patients (<16 years) admitted to our hospital. Our system was based on the model devised by Drake and colleagues [5] but was adapted and extended to include a qualitative assessment of impact on the patient's experience. We present our results following 2 years of data collection, propose three quality indicators that would permit ongoing monitoring of quality and comparison between units and review the current literature on morbidity in paediatric neurosurgical patients.

Methods

The paediatric neurosurgical unit at the Royal Hospital for Children, Glasgow, is the largest unit in Scotland, and in addition to providing neurosurgical care to the population of the west of Scotland (population 2.7 million), it is one of the designated supra-regional national UK centres for craniofacial surgery. The unit has four consultant paediatric neurosurgeons, a specialist trainee, a fellow, an advanced nurse practitioner and a clinical nurse specialist.

The model devised is based on that proposed by Drake and colleagues [5], each adverse event categorised in the scheme outlined in Table 1.

To be included, an adverse event had to occur either during their inpatient stay or if discharged within 30 days of surgery. Wherever possible, the adverse event is linked to the appropriate neurosurgical operation.

Table 1 Classification of complications

Surgical	Neurological deficit	New deficit	
	Meningitis	Excluding indwelling drain	
	Seizures		
	Wound infection		
	CSF leak		
	EVD/lumbar drain infection	Suspected or confirmed	
	Shunt blockage		
	Shunt infection	Suspected or confirmed	
	Post-operative haemorrhage	Managed medically	
	Post-operative haemorrhage	Returned to theatre	
	Post-operative infarction		
	Other		
	Medical	Cardiac	
		Respiratory	Inc pneumonia
GI/hepatic			
Renal/GU/UTI			
Haematological/thromboembolic			
Metabolic			
Cognitive			
Other			

Each potential adverse event is prospectively recorded and then discussed at a monthly review meeting (chaired by the unit clinical governance lead, EC). The adverse event is assigned a category and linked to the appropriate operation. An additional patient impact score was assigned using our organisation's matrix for severity categorisation of complications [11] (Table 2).

If the adverse event resulted in the patient returning to theatre for a further procedure, the impact score was a minimum of 3. An adverse event with an impact score 3, 4 or 5 was termed significant.

A governance meeting, attended by the entire paediatric neurosurgical unit, with representation from allied medical and surgical specialties is held every quarter. At this meeting, each event is reviewed and the final decision whether it is to be included in the morbidity database, its category and impact score is agreed. All inpatient deaths are also reviewed. Multiple complications in the same patient were counted separately, and each was linked to the appropriate procedure, if the patient had undergone more than one procedure.

Three key indicators were identified to permit monitoring of quality of care delivered:

1. Significant adverse event rate—the proportion of operations to which at least one significant adverse event was linked to.
2. Unscheduled return to theatre rate—the proportion of operations which subsequently required an unplanned return to theatre due to a recorded significant adverse event.
3. Surgical site infection rate—the proportion of operations that were linked to any adverse event affecting the surgical site (wound, shunt or drain infections).

The database was recorded and analysed on Microsoft Excel, and chi-squared test was used to analyse categorical variables and a two-sample *t* test to analyse continuous variables. The project was approved by the local clinical governance support unit.

Results

Data was prospectively collected from 1 January 2014 until the 31 December 2015 for all patient aged <16 years, admitted under the care of the paediatric neurosurgical team. Five hundred forty-nine procedures were performed in 287 patients (163 males, 124 females). The mean age at the time of the operation was 5.8 ± 5.0 years (mean \pm standard deviation). One hundred thirty-five (24.6%) operations were performed in patients less than 1 year of age at the time of the operation.

One hundred thirty-eight (25.1%) operations were classified as routine; the remainder was non-elective (urgent and emergency procedures). The principal surgeon was the

Table 2 NHS Greater Glasgow and Clyde impact/severity descriptors

1	Negligible	Reduced quality of patient experience/clinical outcome not directly related to delivery of clinical care
2	Minor	Unsatisfactory patient experience/clinical outcome related to care provision—readily resolvable
3	Moderate	Unsatisfactory patient experience/clinical outcome; short term effects—expect recovery <1 week
4	Major	Unsatisfactory patient experience/clinical outcome; long-term effects—expect recovery >1 week
5	Extreme	Unsatisfactory patient experience/clinical outcome; continued ongoing long-term effects

consultant paediatric neurosurgeon in 307 procedures (55.8%); the majority of the remainder of cases were performed by a trainee as the principal surgeon, supervised by the consultant paediatric neurosurgeon (228 procedures, 41.5%). A small minority of cases were performed by an adult consultant neurosurgeon (14 procedures).

Eight patients died during the study period, six of whom had undergone a neurosurgical procedure in our department during their last hospital stay. All deaths were formally reviewed at the quarterly governance meeting. In the review of two of the deaths, it was concluded that given the underlying diagnosis, the death was unexpected and the adverse events that had occurred during the final inpatient stay were likely to have contributed to the patient's death. One patient (31 days old) suffered an intraventricular haemorrhage following embolisation of a vein of Galen malformation, an external

ventricular drain was inserted, but the baby never recovered from the haemorrhage, and treatment was withdrawn. The second death occurred in a 2-year 3-month-old child who presented in coma with a posterior fossa tumour. Despite emergency surgery and excision of the tumour, she suffered a devastating hypoxic ischaemic brain injury and treatment was ultimately withdrawn; histopathology showed the tumour to be a pilocytic astrocytoma.

For the remaining six deaths, it was concluded that the death was expected and the result of the underlying disease process rather than any adverse event that had occurred.

The calculated crude mortality rate for patients undergoing neurosurgery in our department was 6/287 = 2.1%.

One hundred ninety adverse events were recorded; 130 had an impact score of 3 or greater and were termed significant. Table 3 outlines the category and impact score of the recorded

Table 3 Frequency of adverse event by category and impact score

		Impact score, no. of events					Total	
		1	2	3	4	5		
Surgical	Neurological deficit		3	3	12	1	19	
	Seizures		5	1	2		8	
	Wound infection		2	6	1		9	
	Wound breakdown		1	1	2		4	
	CSF leak		20	21			41	
	EVD/lumbar drain infection		2	6	1		9	
	Shunt blockage				27	3	30	
	Shunt infection			7	4	1	12	
	Post-operative haemorrhage (medical)		1	1			2	
	Post-operative haemorrhage (surgical)					1	1	2
	Other ^a	4	7	8	3	1	23	
	Medical	Cardiac						0
Respiratory				3	3		6	
GI/Hepatic			1				1	
Renal/GU/UTI				1			1	
Haematological/thromboembolic					1		1	
Metabolic		1	11	2	3		17	
Other			2	2		1	5	
Total		5	55	89	36	5	190	

^a The 12 surgical, significant adverse events (impact score 3, 4 or 5) recorded under 'other' included a delay in treatment of raised ICP (1), injury to the small bowel during insertion of a ventriculoperitoneal shunt (1), pressure sore (2), EVD blocked (3), dislocated right elbow (1), ICP monitor or vascular long line dislodged (2) and failure to drain all ventricles following revision of VP shunt (2)

adverse events. Thirty-one adverse events were medical, and 158 adverse events were surgical. The commonest adverse event was a CSF leak; the commonest significant adverse event was a shunt blockage.

Significant adverse event rate

One hundred eleven operations were linked to at least one significant adverse event, giving a significant adverse event rate of 20.2% (6/190 adverse events were not be linked to a neurosurgical procedure).

Unscheduled return to theatre rate

Of the 549 operations performed, 81 were associated with an adverse event that resulted in an unscheduled return to theatre, 14.8%.

Surgical site infection rate

Thirty operations were associated with a surgical site infection or a suspected infection for which treatment was instigated; the overall surgical site infection rate was 5.5% (30/549). The causative organism was identified in 23 of the 29 infections; 26 of the infections were classified as a significant adverse event.

CSF diversion operations had a higher significant adverse event rate (26.5%) compared to spinal operations and all other cranial procedures (21.9 and 12.2%, respectively) ($p < 0.001$). Within the subcategories of cranial surgery, tumour surgery had the highest significant adverse event rate, 32.1% (Table 4).

Routine operations had a lower significant adverse event rate than non-routine operations (11.6 vs 23.1%, $p = 0.004$).

There was no real difference in the significant adverse event rate in operations where the paediatric consultant was the principal surgeon and where they were not (20.5 vs 19.8%, $p = 0.84$). There was no statistical difference in the proportion of male and female patients who experienced a significant

adverse event following surgery (male 21.4 vs 18.6%, $p = 0.42$); nor was there any difference in the mean age of patients who experienced a significant adverse event (6.23 vs 5.71 years, $p = 0.33$).

Discussion

In his recent book, 'Black Box Thinking' [17], Syed explores how success happens and analyses the approach that different industries and professions take when failure occurs. He draws many parallels between the aviation industry and healthcare; both are safety-critical, high-performance professions, which utilise increasingly complex systems, but he argues that there is a profound difference in how they approach adverse or unexpected events. Pilots and aviation system experts perceive such events as the consequence of the conflict between the complexity of the system and their capacity to understand it. In healthcare, Syed contends that too often, such events are seen not as the inevitable consequence of complexity but as an indictment of those who make them. Our development of a surgical morbidity surveillance system is the first step in this process of changing the culture to adverse events in our department.

What is a surgical complication?

Dindo and Clavien [3] emphasised the distinction between the failure to achieve the surgical goal and a complication in their definition; "any deviation from the ideal post-operative course that is not inherent in the procedure and does not comprise a failure to cure."

Sokol and Wilson [16] differed and argued that an undesired outcome, which would include a failure to cure, is perceived as a surgical complication; "any undesirable, unintended and direct result of an operation affecting the patient, which would not have occurred had the operation gone as well as could have been reasonably be hoped".

Table 4 Category of operation

Category of operation	Number of operations	Number of operations linked to significant adverse event	Significant adverse event rate (%)
Brain tumour	53	17	32.1
Epilepsy	8	0	0
Trauma	16	0	0
Craniofacial	48	3	6.3
Vascular	22	3	13.6
Other	83	5	5.6
All cranial	230	28	12.2
CSF diversion	287	76	26.5
Spine	32	7	21.9

If the goal of a prospective morbidity surveillance programme is to identify areas where surgical care could be improved, we argue that this should include when the primary goal of the operation has not been achieved. We sought to identify any adverse event that had occurred in patients admitted under our care and thus adopted the Sokol and Wilson definition that this would mean any clinical event that was undesired and unintended.

Classification of surgical complications

The Clavien and Dindo classification [4] is the most widely cited classification system; designed for general surgery, its application has been extended to related surgical specialties, for example to urology, and Ibanez et al. [6] proposed a modified form of the system for neurosurgical patients. The Clavien-Dindo system classifies a complication according to the therapy used to treat it and thus is an assessment of impact severity; for example, wound infection requiring surgical washout and replacement of an external ventricular drain would both be grade IIb: complication requiring intervention with general anaesthesia. As it does not make a distinction between different categories of complication, its practical use as a means for improving clinical practice is more limited.

It is also important to remember that these systems were designed for adult patients; taking action after an adverse event in a child may require a greater level of intervention than in an adult; for example, resuturing a wound or performing a lumbar puncture may require a general anaesthetic in a child, but this is less likely in an adult; thus, the grade of the complication will have increased.

We adopted the system proposed by Drake and colleagues [5] for several reasons. Firstly, it was specifically designed for neurosurgical patients and the categorisation of the complication by surgical and medical subgroups would be easily applied in a busy paediatric neurosurgical unit. The same complication occurring with different procedures could more easily be identified. Secondly, the system has been adopted by other paediatric neurosurgical units [19], permitting a comparison between units. Lastly by linking each complication to an operation, the Drake system creates a simple and easily counted denominator. This gives a more accurate assessment of the significant adverse event rate than linking it to individual patients; for example, estimating the 30-day shunt blockage rate is more meaningful if we compare the number of shunts that blocked in 30 days to the total number of shunts inserted, rather than the total number of patients who underwent the procedure.

We modified the model to include an assessment of patient impact severity, using our organisation's clinical risk matrix. Drake et al. [5] classified morbidity "as any significant

adverse outcome or death"; we felt that the term significantly needed to be more precisely defined. Whilst our system is simpler than the Clavien-Dindo system, we believe that the dual categorisation of an event adds strength.

Comparison of complication rates

Our significant adverse event rate was 20.2%; in comparison to other units using the same system, Drake et al. [5] reported an overall complication rate of 16.4% over 2 years and van Lindert et al. [19] reported an overall complication rate of 20.2%. Using a different system, Patel et al. [13] also reviewed all cranial and spinal surgery and published a 30-day morbidity rate of 15.0%. Taking into account differences in interpretation and data collection, we believe that our result that approximately one in five neurosurgical procedures resulted in an adverse event is in keeping with current published rates for paediatric neurosurgical units.

Moiyadi et al. [9] and Neervoort et al. [10] reviewed the morbidity associated with cranial operations for CNS tumours and identified morbidity rates of 44.4 and 68.6%, respectively. We found that our cranial operations for tumour have a significant adverse event rate of 32.1%; this trend of higher morbidity for tumour operations in comparison to the overall morbidity rate within a unit was also recorded by van Lindert [19], 32.7%, and Drake [5], 27.9%. This study supports the view that morbidity following tumour surgery is greater than other types of neurosurgery in paediatric patients.

In reviewing our significant adverse event rates, we identified that our significant adverse event rate (SAER) was greater for CSF diversion procedures, 26.5% (shunt insertion and revision, external ventricular drain insertion, endoscopic procedures and open craniotomy procedures for arachnoid cysts), than for other cranial procedures or spinal procedures. This appeared to be higher than that published by other units (van Lindert et al. [19] 21.8% and Drake et al. [5] 15.4%). In light of this finding, a detailed analysis of the CSF diversion procedures was completed. It was found that the SAER was highest for shunt procedures (71 new shunt insertions and 75 shunt revisions) and the commonest surgical complication was shunt blockage. Whilst it appeared that during the 2-year period of the study, we had at least three complex patients in whom it proved extremely difficult to maintain a working CSF drainage system, our department identified several areas where intraoperative technique could be modified with the aim of reducing the frequency of early shunt failure.

We reported an unscheduled (30 days) return to theatre rate of 14.8%; this excluded those patients who had a planned return to theatre for a planned second operation, for example tumour resection after a first operation to treat hydrocephalus.

A rate of 14.8% is in keeping with the rates published by other paediatric neurosurgical units; Van Lindert and colleagues [19] reported their unscheduled return to theatre rate

Table 5 Literature review of published morbidity and mortality studies in paediatric neurosurgical patients

Study	Age of patients (years)	Duration of observation (days)	Type of surgery	Number of patients	Number of procedures	Morbidity (%)	Mortality (%)	30-day reoperation rate (%)	Surgical site infection rate (%)
Albright [1]	NS	Varies	Shunt insertion/shunt revision/craniotomy for brain tumour/craniotomy for sagittal synostosis/tethered cord	200/72/100/91/100	200/72/100/91/100	NS	1% for brain tumours	NS	7/11/4/0/9, Composite infection rate, 6.6
Smith [14]	0–18	Inpatient stay	Cranial—tumours only	4712	NS	NS	1.60	NS	NS
Drake [5]	NS	30 days	All cranial and spinal surgery	NS	1082	16.4	11 deaths	10.4 (estimated)	3.1 (estimated)
Ncervoort [10]	0–18	NS	Cranial—tumours only	121	137	68.6	0.8	NS	NS
Moyyadi [8]	0–18	NS	Cranial—tumours only	117	117	44.4	7.7	Reexploration, 8.5; additional procedure, 14.5.	15.4
Markerji [9]	NS	300 days	All cranial and spinal surgery	NS	NS	NS	2.7	17.3	NS
Solheim [15]	0–16	30 days	Cranial—tumours only	1242	1242	NS	3.1	NS	NS
Von Lehe [18]	0–16	30 days	Cranial	641	769	2.0	NS	NS	3.9
O’Kane [12]	0–14	30 days	Cranial—tumours only	1280	NS	NS	2.7	NS	NS
Patel [13]	NS	30 days	All cranial and spinal surgery	1990	3195	15.0	1.6	NS	NS
Van Lindert [19]	0–17	30 days	All cranial and spinal surgery	581	1000	20.2	2.2	10.5	3.4
Desai [2]	NS	30 days	Emergency and urgent surgery	580	710	10.3	5.2	4.7	3.3
Van Lindert [20]	0–17	Intraoperative only	All cranial and spinal surgery	1148	1807	3.5	0.2	NS	NS
Current study	0–16	30 days	All cranial and spinal surgery	287	549	20.2	2.1	14.8	5.5 (4.2% organism identified)

NS not specified

of 10.5%; Mukerji and colleagues [9] reported a 30-day unplanned reoperation rate of 17%.

Mukerji et al. advocated the adoption of an unplanned reoperation rate as a quality indicator; we agree with their view that it is an easily counted, common event that is non-discretionary. They reviewed all reoperations in a 2-year period and found that the majority of the reoperations (61%) occurred within 30 days of the original operation (median time between first and unplanned reoperations was 9 days).

The necessity in identifying the causative organism in a surgical site infection has been extensively debated. We argued that if there was a clinical suspicion of an infection, for example, a child with symptoms of meningitis, a fever and a raised white cell count in their CSF following a shunt revision, the failure to identify the causative organism did not delay or defer treatment; therefore, any child who underwent treatment of an infection should be defined as having an infection.

We linked 29 operations to a surgical site infection: 5.3% of all procedures. Interestingly, the causative organism was identified in 22 of these infections, which if we had used the criteria of identification of the causative organism, employed by other units, would have given a surgical site infection rate of 4.0%.

These rates of surgical site infection are in keeping with that published by other units for both all types of neurosurgery and specific subtypes (Table 5).

Conclusion

Whilst most paediatric neurosurgery centres review their morbidity and mortality, we believe that our unit's surveillance model has formalised this process and permits an ongoing method of both quality assurance and quality improvement in the care that we provide for our patients.

The model that we have adopted was specifically designed for paediatric neurosurgery, and our experience has been that it can be implemented into a busy unit with minimal disruption of day-to-day clinical activity.

The proposed three key indicators will provide a means for comparison between units for both overall and subcategories of surgery, thus further promoting quality assurance within our speciality.

Compliance with ethical standards

Conflict of interest On behalf of all authors, the corresponding author states that there is no conflict of interest.

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Comparison of Quality Metrics for Pediatric Shunt Surgery and Proposal of the Negative Shunt Revision Rate

Thomas Beez and Hans-Jakob Steiger

OBJECTIVE: Shunt surgery is common in pediatric neurosurgery and is associated with relevant complication rates. We aimed to compare previously published metrics in a single data set and propose the Negative Shunt Revision Rate (NSRR), defined as proportion of shunt explorations revealing a properly working system, as a new quality metric.

METHODS: Retrospective analysis of our shunt surgery activity in 2015 was performed. Demographic, clinical, and radiologic variables were extracted from electronic medical notes. Surgical Activity Rate, Revision Quotient, 30-day shunt malfunction rate, 90-day global shunt revision rate, Preventable Shunt Revision Rate, and novel NSRR were calculated.

RESULTS: Of 60 shunt operations analyzed, 18 (39%) were new shunt insertions, and 42 (70%) were revisions. Median age was 18 months (range, 0.03–204 months), and main etiologies were posthemorrhagic ($n = 16$; 41%), congenital ($n = 11$; 28%), and tumor-associated ($n = 8$; 21%) hydrocephalus. Within 90 days after index surgery, 13 shunt failures occurred, predominantly owing to proximal failure ($n = 6$; 46%). Surgical Activity Rate was 0.127, Revision Quotient was 2.333, 30-day shunt malfunction rate was 0.166, 90-day global shunt revision rate was 21.7%, and Preventable Shunt Revision Rate was 38.5%. NSRR was 7.1%.

CONCLUSIONS: Our results correlate with published values and offer measurement of quality that can be compared across studies and considered patient-oriented, easily measurable, and potentially modifiable. We

propose NSRR as a new quality metric, covering an aspect of shunt surgery that was not addressed previously.

INTRODUCTION

Surgical procedures have recently become extensively monitored for various quality metrics, with motivations ranging from protecting patients to reducing health care costs. In the United Kingdom, a series of scandals surrounding medical care led to a call for greater transparency, which was answered by mandatory publication of surgeon-specific outcome indicators. Many health care providers have become concerned about the validity and accuracy of the data provided, as erroneous data may threaten surgeons careers and urge them to practice defensive medicine, omitting high-risk operations and refraining from supporting surgical trainees.^{1,3} However, measuring surgical quality clearly reflects a global interest.

As shunt surgery is frequently performed by pediatric neurologic surgeons and the procedure has been associated with various complications, leading to additional morbidity for patients and substantial financial burden for health care systems, shunt surgery would be a suitable surgical procedure for quality and outcome studies.³⁻⁵ Although several studies attempted to identify risk factors for shunt failure and reduce its incidence, no consensus on predictive variables exists to date.^{6,7} Recently, several authors suggested different quality metrics to objectively assess and compare quality of shunt surgery.⁸⁻¹¹ However, no common data elements have been agreed on, and thus comparison of results with benchmarks or across studies remains limited.

A quality metric should be patient-oriented, should make sense, and should be measurable as well as modifiable.^{10,12} Looking beyond neurosurgery, we became aware of the negative

Key words

- Hydrocephalus
- Infection
- Malfunction
- Pediatric
- Quality
- Shunt

Abbreviations and Acronyms

- NSRR: Negative Shunt Revision Rate
- PSRR: Preventable Shunt Revision Rate

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appendectomy rate in general surgery. Negative appendectomy is defined as the nonincidental removal of a normal appendix. It occurs at a rate of up to 15% and has been linked with additional morbidity and health care costs.¹³ Although critics initially insisted that reducing this rate would lead to higher incidence of appendiceal perforation owing to delayed or missed diagnoses, this hypothesis has been proven false.¹⁴ Introduction of computed tomography and clinical algorithms has lowered the negative appendectomy rate to 3%.¹⁵ Despite some criticism, this rational quality metric appears to be directly relevant to the patient, is measurable, and has been demonstrated to be modifiable. In the present study, we sought to compare previously published quality metrics for shunt surgery in children in a single data set. Additionally, we have transferred the concept of the negative appendectomy rate to neurosurgery and propose the Negative Shunt Revision Rate (NSRR) as a new quality metric, defined as the proportion of shunt revisions, more accurately termed explorations, revealing a properly working shunt system.

MATERIALS AND METHODS

Patients

This retrospective analysis was approved by the institutional review board (study number 5949R) and was registered a priori in the institutional study registry (registration number 2017044234). Patients <18 years old who underwent either implantation or revision of a cerebrospinal fluid shunt in 2015 were identified from an institutional electronic database. Age, sex, etiology of hydrocephalus (congenital, posthemorrhagic, tumor, dysraphism, or other), shunt type (ventriculoperitoneal or ventriculoatrial), burr hole position for ventricular catheter insertion (frontal or occipital), and number of previous shunt operations were retrieved from electronic medical notes. The preoperative frontal-to-occipital horn ratio was calculated from axial cranial computed tomography or magnetic resonance imaging as previously described. Shunt

failures within 30 days and 90 days were recorded for each index operation, both new implantations and revisions. Type of shunt failure was classified as proximal failure, valve failure, distal failure, infection, or wound revision. The exact etiology of each failure was determined from operative notes, and preventable failures were defined as proposed by Venable et al.¹⁰ Infection was defined by suggestive clinical signs (e.g., fever, symptoms of meningitis, cerebrospinal fluid pleocytosis, signs of infection at shunt wounds or along the shunt tract) with or without positive microbiologic cultures.

Shunt Quality Metrics

The Surgical Activity Rate, Revision Quotient, 30-day shunt malfunction rate, 90-day global shunt revision rate, and Preventable Shunt Revision Rate (PSRR) were calculated for our data set according to the definitions summarized in Table 1.^{8–11} The novel NSRR was defined as the proportion of shunt revisions per year revealing a properly working shunt system in relation to the overall number of annual shunt revisions. To fulfill the criteria of a negative shunt revision, the operation notes had to explicitly state that no intraoperative malfunction had been identified, resulting in either leaving the original shunt in situ or revising the functioning shunt and/or components for prophylactic reasons.

Statistical Analysis

We performed descriptive statistics, and all variables are presented as medians and range or absolute numbers and percentages, unless otherwise specified. Data were analyzed with GraphPad Prism 3.00 (GraphPad Software, Inc., La Jolla, California, USA).

RESULTS

Patients

For the year 2015, 39 children undergoing 60 shunt operations were identified from our institutional database. The median age of this cohort was 18 months (range, 0.03–204 months); there were

Table 1. Definitions of Shunt Quality Metrics

Reference	Quality Metric	Definition	Interpretation/Range
Barton et al., 2013 ⁸	SAR	Number of definitive operations for treatment of hydrocephalus per year divided by number of patients with hydrocephalus seen in follow-up during that year	0.115–0.299 in simulated neurosurgical practices
Barton et al., 2013 ⁸ ; Platt and Freibott, 2014 ⁹	RQ	Number of definitive revisions per year divided by number of definitive operations for newly diagnosed hydrocephalus during that year	Median RQ 2.53 (range, 0.06–10.92) in KID
Platt and Freibott, 2014 ⁹	30-day shunt malfunction rate	Number of initial VP shunt placement procedures performed on children 0–18 years old that malfunction and result in shunt revision within 30 days of initial placement divided by total number of initial cerebrospinal VP shunt procedures performed on children 0–18 years old	13.5% of all shunts in NSQIP-P database
Venable et al., 2016 ¹⁰	PSRR	Preventable failure within 90 days defined as failure subsequent to infection, malposition of proximal or distal catheter, or improperly assembled or inadequately secured shunt resulting in migration, disconnection, or kinking	33% (of 90-day failures); 80.6% of these preventable failures occurred within first 30 days
Rossi et al., 2016 ¹¹	90-day raw or global shunt revision rate	Percentage of shunts failing within 90 days from index surgery defined as implantation of new shunt or revision or augmentation of existing shunt system	24.1%

SAR, Surgical Activity Rate; RQ, Revision Quotient; KID, Kids Inpatient Database; VP, ventriculoperitoneal; NSQIP-P, National Surgical Quality Improvement Program Pediatrics; PSRR, Preventable Shunt Revision Rate.

19 female and 20 male patients. The main etiologies were posthemorrhagic ($n = 16$; 41%), congenital ($n = 11$; 28%), and tumor-associated ($n = 8$; 21%) hydrocephalus, with other causes in 4 patients (10%). The mean preoperative frontal-to-occipital horn ratio was 0.42 (range, 0.32–0.60).

Shunt Operations

This study included 60 shunt operations comprising 18 (39%) new shunt insertions and 42 (70%) shunt revisions. A ventriculoperitoneal shunt was inserted in 55 cases (92%), and a ventriculoatrial shunt was inserted in 5 cases (8%). Ventricular catheters were inserted through either an occipital ($n = 41$; 68%) or a frontal ($n = 19$; 32%) burr hole.

There were 13 shunt failures within 90 days after index surgery, with 7 of these failures occurring in the first 30 days. The mean time to failure was 28 days (range, 2–86 days). The index operations were 5 new insertions and 8 revisions. In 6 cases, a proximal failure was found, with misplacement of the ventricular catheter in 3 cases and obstruction in 3 cases. Distal shunt failure occurred in 2 cases, with the underlying causes being obstruction of the distal catheter and peritoneal malabsorption, respectively. The remaining shunt failures were due to wound breakdown ($n = 2$), shunt infection ($n = 2$), and valve obstruction ($n = 1$).

Shunt Quality Metrics

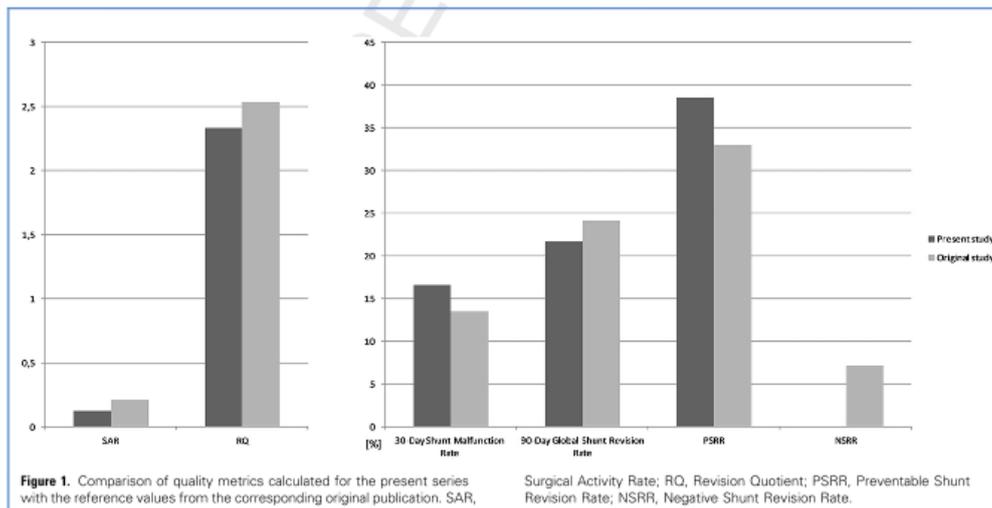
For calculation of the Surgical Activity Rate, the number of patients with hydrocephalus seen in follow-up during 2015 was retrieved from the electronic hospital documentation and determined to be 142. Thus, for this cohort, the Surgical Activity Rate was 0.127. With 42 shunt revisions and 18 new shunt insertions,

the Revision Quotient for this series was 2.333. Of the 7 shunt failures within 30 days, only 3 occurred after initial implantation of a ventriculoperitoneal shunt (i.e., after a new insertion), leading to a 30-day shunt malfunction rate of 0.166 or 16.7%. The 90-day global shunt revision rate was 21.7%. As 5 of the 13 shunt failures occurring within 90 days after index surgery were considered preventable according to the criteria defined by Venable et al.,¹⁰ the PSRR was calculated as 38.5%.

Of the 42 shunt revisions in this series, 3 were considered negative shunt revisions, as no shunt malfunction could be detected intraoperatively despite clinical suspicion of shunt failure. Therefore, the novel NSRR was 7.1%. One patient presented with a tense fontanel, 1 patient presented with vomiting and a slowly refilling shunt reservoir, and the third patient presented with only a slowly refilling reservoir. Cranial imaging with regard to ventricular size was unchanged in all 3 patients. The functioning shunt was left in situ in 2 patients and was completely revised in 1 patient. The quality metrics calculated for the present series are compared with the reference values from the literature in Figure 1.

DISCUSSION

Shunt surgery for hydrocephalus is a major part of a pediatric neurosurgeon's routine workload and a field of intense clinical research.^{5,10} However, shunts continue to have relatively high failure rates and can be associated with considerable morbidity for individual patients and significant costs for health care systems.³ Several studies have attempted to identify risk factors for shunt failure, but most risk factors are not unequivocally reproducible across studies and are thus considered inconsistent and



controversial.^{6,7} Research efforts worldwide are directed toward generating scientific evidence. The North American Hydrocephalus Clinical Research Network serves as a model of collaborative multicenter research in this field, with several finished and ongoing studies covering aspects of pediatric shunts from ventricular catheter entry site, risk factor analysis, outcome assessment, and validation of protocols for reduction of failure.^{6,17-22} In an effort to measure quality, previous authors have published proposals ranging from the relatively raw global shunt revision rate to metrics focusing on particular aspects, such as the PSRR.^{10,11} However, in the absence of established reference ranges, such metrics offer limited comparison and insight, especially for patients, and might simply amount to intellectual exercises.¹² Nevertheless, the relevance of quality metrics remains high in times of accountable medicine. In the United Kingdom, this development recently culminated in the government-mandated publication of named surgeon-specific outcome data.¹ As critics argue that this initiative relies on databases and metrics that were not designed for such purposes, efforts to measure quality in pediatric hydrocephalus might be foresight rather than just intellectual exercises.

We undertook the present study to determine established quality measures for pediatric shunt surgery within our own patient population at a small academic pediatric neurosurgery unit. The main finding was that all metrics could be calculated with limited additional effort, and the results fall closely within the range suggested in the original studies. Therefore, in our opinion, these quality metrics are practical, reproducible, and comparable.

Previous authors have suggested that quality metrics should be measurable and modifiable, make sense, and be patient-centered.^{10,12} Although the first 3 attributes are fulfilled by most metrics, the fourth is critical. One could argue that an affected patient experiences a preventable failure similar to any other unplanned revision surgery. However, a low PSRR would reflect good performance in this aspect of shunt surgery and thus make sense to patients and physicians alike. As there are certainly multiple variables influencing quality of shunt surgery, we hypothesized that, with the exception of very raw measures, capturing all aspects within 1 metric is difficult. The disadvantage of such raw measures is the difficulty to derive focused quality improvement efforts. However, such efforts start with a systematic database ensuring relevant common data elements, as suggested for shunts within the National Surgical Quality Improvement Program Pediatrics in North America.^{23,24}

Therefore, we proposed a new quality metric capturing the aspect of shunt revisions or explorations revealing a properly working shunt system. Although some of these operations might be unavoidable in clinical routine, a higher number of shunt explorations owing to migraine headaches or vomiting related to viral gastroenteritis should in our opinion prompt a review of clinical decision making. This approach has been established in general surgery as the negative appendectomy rate. A negative appendectomy is defined as the nonincidental removal of a normal appendix. Despite initial criticism that lowering this rate would lead to a higher incidence of appendiceal perforation owing to delayed or missed diagnoses, this hypothesis has been proven false, and refinement of diagnostic approaches led to a decrease of the negative appendectomy rate from 15% to 3%.^{13,15,25,26} As this

quality metric appears to be directly relevant to the patient, is measurable, and has been demonstrated to be modifiable, we sought to transfer this concept to shunt surgery. In the present study, we determined the novel NSRR to be 7.1% in our cohort. Negative shunt revisions were performed in 3 patients, who presented with red flag signs, including tense fontanel, vomiting, and slowly refilling shunt reservoir. Although cranial imaging with regard to ventricular size was unchanged in all 3 patients, shunt explorations were performed, and properly functioning shunts were found; shunts were left in situ in 2 patients, and the shunt was completely revised in 1 patient. Although we acknowledge that in such inconclusive clinical scenarios a shunt exploration is justified, a high NSRR should lead to a careful review of individual cases and could potentially offer guidance for focused quality improvement. Measures aiming at reduction of a high NSRR might be improvements in diagnostic algorithms for questionable candidates for shunt revision (i.e., children with insufficient proof of shunt failure based on clinical and radiologic criteria alone), such as implementation of ultrasound measurement of optic nerve sheath diameter or invasive intracranial pressure monitoring.²⁷⁻³¹ However, we assume that a single quality metric will always have considerable limitations and that the most accurate capture of quality will be achieved by combining several metrics.

In contrast to the negative appendectomy rate, which relies on objective pathologic analysis of the removed appendix, the NSRR is based on the intraoperative assessment of shunt function by the neurosurgeon performing the revision surgery. Such subjective judgment potentially introduces bias into the calculation of the NSRR. However, we assume that most refined shunt quality metrics rely on a certain degree of subjective judgment. The PSRR relies on the correct classification of the exact reason for shunt failure, which is made by the surgeon. From this point of view, raw global quality measures are probably least susceptible to bias and subjectivity, although their potential yield in guiding precise quality improvement efforts is limited. A further limitation of the present study is inherent in its aim; as we sought to determine established quality measures for pediatric shunt surgery within our own patient population at a small academic pediatric neurosurgery unit, we present single-center, retrospective data with a limited number of patients. Therefore, we cannot provide a reference range for the NSRR or an ideal NSRR. We hope that our contribution to the debate on shunt quality metrics will lead to reproduction and external validation of the NSRR from larger series, multicenter registries, or clinical research networks.

CONCLUSIONS

The quality metrics obtained for our data set correlate with the published values and, analyzed in conjunction, offer a measurement of quality that 1) can be compared across studies using these common metrics and 2) can be considered patient-oriented, easily measurable, and potentially modifiable. Additionally, we propose the NSRR, which focuses on the rate of shunt explorations revealing intact shunt systems. Although careful review is required on a case-by-case basis, a high NSRR could reflect an exceptionally low threshold to revise shunts and might prompt a review of clinical decision making. However, when judging such results, it must be considered that shunt revisions are not exclusively linked

to suboptimal medical care but are partially intrinsic to the disease and procedure and thus to a certain degree beyond a neurosurgeon's control. A particular quality metric seen in isolation will

always have certain limitations, and the most accurate capture of quality will be achieved by combining multiple metrics, including, we hope, the NSRR after external validation.

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Headache and Shunt-Related Impact on Activities of Daily Life in Patients Growing Up with a Ventriculoperitoneal Shunt

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J Neurol Surg A

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Abstract

Background/Aims Ventriculoperitoneal shunts (VPS) are a mainstay of treatment for hydrocephalus. Morbidity related to infection and dysfunction is well known, whereas data on psychosocial outcome and quality of life are scarce. Our aim was to assess headache burden and shunt-related impact on daily life in children growing up with a VPS.

Methods Patients between 3 and 21 years of age were identified and their families were contacted. A set of standardized questionnaires was used to assess headache and quality of life.

Results Fifteen patients with a mean age of 12 years agreed to participate in this study. Satisfaction with the VPS and improvement of the patient's condition was reported by 87%. A total of 67% denied negative impact on everyday life, reported a headache less than once a month, and never or rarely miss school. However, 53% take precautions before special activities.

Conclusions The results demonstrate that modern VPS systems are tolerated very well and do not per se reduce quality of life. However, patients adapt to the presence of a VPS by avoiding risks and taking precautions. Chronic headache is not a major problem and not necessarily attributed to the VPS. Nevertheless, most patients asked for technical improvements, which might guide future research and the technical development of VPS.

Keywords

- ▶ hydrocephalus
- ▶ ventriculoperitoneal shunt
- ▶ outcome
- ▶ quality of life
- ▶ headache

Introduction

A ventriculoperitoneal shunt (VPS) is a common form of treatment for hydrocephalus in children.¹ Most young children receiving a VPS remain shunt dependent throughout their childhood, adolescence, and probably adult life.² Although the impact of morbidity related to shunt infection and dysfunction has been studied extensively, data on psychosocial outcome and quality of life are limited.^{3,4}

However, factors that potentially exert a detrimental long-term influence on quality of life have been identified in VPS patients. For instance, even in the absence of overt VPS-related problems, headache has a higher incidence in patients with VPS compared with the normal population.⁵ The earlier in life a

patient receives a VPS, the greater the influence of such factors might be on all dimensions of development including cognitive, social, academic, and professional outcome.

Our aim was to assess headache symptom burden and shunt-related impact on activities of daily life in children and adolescents growing up with a VPS.

Methods

Patient Cohort

A previous publication from our institution defined a pediatric VPS patient cohort by retrospectively identifying all neurosurgical patients < 16 years of age who underwent

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primary implantation of a VPS between January 2005 and December 2010. From this cohort, patients aged between 3 years (preschoolers) and 21 years (transition into adult neurosurgical care) with at least 1-year follow-up since initial VPS implantation at the time of the present study were identified, and families were contacted by phone or mail whether they would participate either by interview or written questionnaire.

Methodology

A German-language test battery was designed that consisted of two parts. Part A was a nonstandardized 7-item questionnaire exploring the subjective satisfaction with the VPS in terms of symptom control, impact on everyday life, impact on special activities, and technical aspects. Part B was a standardized German-language questionnaire for chronic headache in childhood (Structured Headache Interview for Children, or Strukturisiertes Kopfschmerzinterview für Kinder [SIKI]).⁶ This structured interview explores both the child's as well as the parents' perception of headache symptoms, triggers, coping mechanisms, emotions, and a subjective disease model.

Results were analyzed using descriptive statistics. Approval from the local medical ethics committee was obtained (study 4212).

Results

Patients

Forty patients were contacted, and 15 patients responded and agreed to participate in this study. The mean age at the time of interview was 12 years (range: 3–21 years). The mean follow-up time since initial VPS implantation was 7 years (range: 2–18 years). Nine patients were male; six were female. Indications for initial VPS insertion were congenital hydrocephalus (*N* = 8), tumor-related hydrocephalus (*N* = 4), and posthemorrhagic hydrocephalus (*N* = 3). The

mean age at initial VPS insertion was 2 years (range: 0–15 years). The most common valve types at the initial operation were Codman Hakim differential pressure (*N* = 5) and paedigAV gravitational (*N* = 4) valves. Before the time of interview, 12 VPS revisions had been performed in 9 patients, with 10 revisions secondary to mechanical shunt failure and 2 revisions because of infection.

Part A: Nonstandardized 7-Item Questionnaire

Fig. 1 summarizes the main results from this questionnaire. Overall satisfaction with the VPS (item 1) and improvement of the patient's condition (item 2) were confirmed by 13 responders (87%), respectively. Item 1 allowed for further free-text answers. A 17-year-old young man reported being teased by schoolmates because of his visible shunt tubing, but he denied any physical constraints due to the VPS. A 21-year-old woman complained of visible shunt tubing at the chest level, and for this reason she avoids wearing clothes with a low-cut neckline. Two responders specifically appreciated the easy programmability of their Codman Hakim valves.

Ten responders (67%) denied a negative impact of the VPS on everyday life (item 3), and 11 (73%) consider the VPS reliable (item 4). Item 3 allowed for free-text answers. Five responders uniformly described subjective limitations with regard to sports (gymnastics, water sports, cycling, football) and/or precautions to avoid bumping or touching the valve (additional caution, e.g., in nursery/school, at the hairdresser, during sports).

Eight responders (53%) take precautions before special activities (item 5), such as traveling, whereas seven (47%) do not. Free-text answers for this item uniformly described precautions while traveling: sufficient hydration in hot weather (*N* = 2), additional sun protection (*N* = 2), carrying the shunt valve patient ID card (*N* = 3) and relevant medical notes (*N* = 2), and collecting information about the neurosurgical unit nearest to the holiday location before departure (*N* = 2).

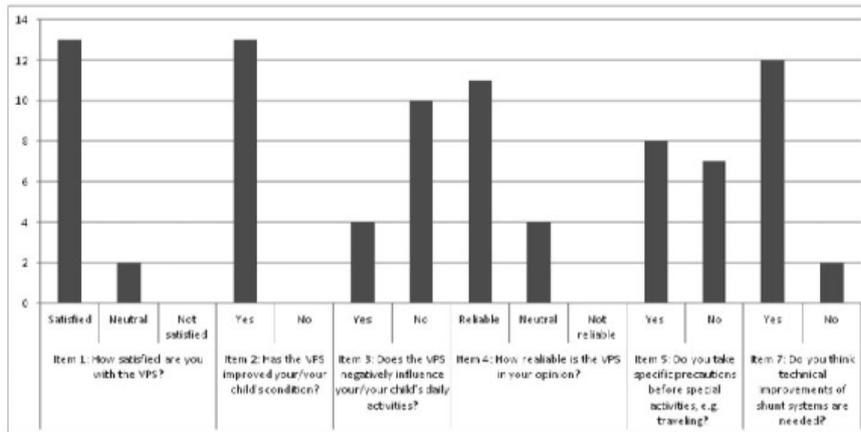


Fig. 1 Main results from part A (nonstandardized 7 item questionnaire). VPS, ventriculoperitoneal shunt.

Most of the responders ($N = 12$ [80%]) asked for technical improvements of the contemporary VPS systems, whereas only two (13%) considered it technologically mature (item 7). Item 7 allowed for free-text answers. Responders asked for smaller and less visible VPS tubing and valves ($N = 5$) and improved durability ($N = 3$).

Part B: Standardized Headache Questionnaire

Fig. 2 summarizes the main results from part B of the questionnaire. Ten responders (67%) reported headache less than once a month and three (20%) every week (item 8). Very short episodes of headache were described by four (27%) and episodes of ~ 12 hours were reported by six (40%) of the responders (item 9). In the majority ($N = 6$ [40%]) headache have a slow onset (item 10). Headaches are present when waking up in three (20%), occur during morning or at noon in three (20%), or during the afternoon and evening in four (27%) cases, whereas six patients (40%) did not notice a pattern (item 11). The prevailing location of headaches (item 12) in this cohort was the frontal area ($N = 12$ [80%]). Headache character (items 13–20) was described as oppressive ($N = 7$ [47%]), sharp ($N = 3$ [20%]), or throbbing ($N = 3$ [20%]). The mean pain intensity on the visual analog scale (item 21) was 5 (range: 0–8). Concerning triggers of headache (items 34–51), eight responders reported sleep deficit as an occasional or frequent trigger, seven (47%) changes in climate or weather, five (33%) light, four (27%) noise, four (27%) rush and time pressure, three (20%) physical overexertion, three (20%) tension, and three (20%) problems at school.

Regarding impact on activities of daily life (items 65–72), 10 patients (67%) never or rarely have to miss school, and 7 patients (47%) never or rarely have to leave homework undone. Ten responders (67%) are never or rarely prevented from meeting friends and performing leisure activities, respectively.

For patients, the subjective main reason for headaches (item 75) was the VPS itself ($N = 3$ [20%]), dehydration ($N = 2$ [20%]), neck tension ($N = 2$ [13%]), or climate ($N = 2$ [13%]). For parents, the subjective main reason for their child's headaches (item 15) was dehydration ($N = 4$ [27%]), common cold ($N = 3$ [20%]), and sleep deficit ($N = 3$ [20%]). Parents considered their child's headache frequency stable or improving in 9 (60%) cases and the intensity stable or improving in 10 cases (67%).

Discussion

This study demonstrated that most young patients growing up with a VPS report overall satisfaction with the system, experienced a significant improvement of their symptoms, and consider the VPS a reliable treatment. Although most patients deny significant limitations due to the VPS in everyday life, a large number avoid higher risk activities, such as certain sports. More than 50% of patients and families take special precautions when going on vacation, mainly related to preparedness for a medical emergency abroad (carrying the shunt valve patient ID card, relevant medical notes, and collecting information about the neurosurgical unit nearest to the holiday location).

Previous studies enrolling similar cohorts (i.e., children and adolescents treated with VPS) evaluated quality of life with the 36-Item Short Form Health Survey (SF-36) and Barthel Index⁷ or the Hydrocephalus Outcome Questionnaire (HOQ) and Health Utilities Index Mark 3 (HUI3).⁸ The first study concluded that self-perceived health was poorer as assessed by SF-36 as compared with the background population. The latter study found a mean HOQ Overall Health score of 0.68, with 0 the worst and 1 the best quality of life. Interestingly, this study identified shunt complications as a major determinant of quality of life, which is a modifiable risk factor. This notion

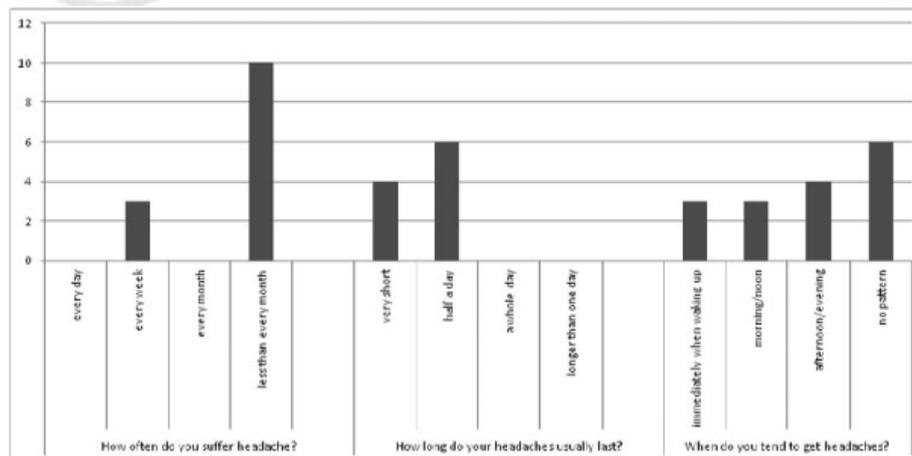


Fig. 2 Main results from part B (standardized German-language questionnaire for chronic headache in childhood: Structured headache interview for children, or Strukturiertes Kopfschmerzinterview für Kinder [SIKI]).⁶

is supported by most of our patients, who asked for technical improvements of the shunt system in the future with regard to durability, although the overall reliability of contemporary VPS systems was rated positive in our cohort. However, another study used HOQ to compare quality of life in patients treated with VPS versus endoscopic third ventriculostomy (ETV) and did not find any substantial difference in quality-of-life outcome between these two treatment options for hydrocephalus after multivariable adjustment.⁹ These results might suggest that the underlying etiology of hydrocephalus could be more relevant for outcome than the actual treatment modality, as shown by Hommet et al.¹⁰

Headache was relatively uncommon in our cohort, with 70% experiencing headache less than once a month and only 20% once a week. Headaches tend to be frontal, with an oppressive character and medium intensity. Common triggers of headache were sleep deficit and changes in climate or weather. Concerning impact on everyday life, 70% of patients never or rarely miss school, cancel meetings with friends, or refrain from leisure activities due to headache. Interestingly, the vast majority of patients and parents do not attribute headaches to the VPS or the underlying hydrocephalic condition. Most patients have a stable or improving headache symptom burden while growing up with their VPS.

Although some authors conclude that patients with VPS have a higher incidence of severe headaches than normal populations,⁵ others suggest that the number of VPS patients with chronic headache unrelated to modifiable shunt problems and requiring specialist pain management is actually very low.¹¹ However, as supported by our results, there is certainly a subgroup of VPS patients with chronic headache that is idiopathic rather than shunt related. However, before making this diagnosis it is mandatory to exclude modifiable shunt problems such as underdrainage, overdrainage, and slit ventricle syndrome.^{11,12}

The main limitation of the present study is the relatively small patient population that does not allow statistical analysis with regard to differences in headache incidence or quality of life between subgroups (e.g., with different VPS valves, different etiology of hydrocephalus, or number of shunt-related complications or revisions). The response rate of 38% leads to a certain nonresponse bias but is still within the expected range according to previous analyses of questionnaire methodology¹³. Additionally, direct comparison with normal subjects or ETV patients would be interesting.

Conclusions

We consider this study to reflect realistic clinical practice because we explored our patients' experience with a

tangible approach yielding meaningful results. With regard to medical care and counseling of young VPS patients and their families, the main implications of this study are (1) modern VPS systems are tolerated very well and do not per se reduce quality of life, (2) patients and families nevertheless adapt to the presence of a VPS by avoiding risks and taking additional precautions, and (3) chronic headache is not a major problem and not necessarily attributed to the VPS. However, 80% of our patients asked for technical improvements in terms of smaller size and higher durability, which might guide the direction of future research and technical development of VPS.

Conflicts of Interests Disclosure

The authors report no conflicts of interest.

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