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Medical and Socioeconomic Predictors of Psychosocial Functioning in Pediatric Hydrocephalus

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ABSTRACT

Medical and Socioeconomic Predictors of Psychosocial Functioning in Pediatric Hydrocephalus

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Hydrocephalus can impact all areas of health, including physical, cognitive, and psychosocial functioning. Etiology can be a major factor in health outcomes, but prior research on psychosocial functioning in hydrocephalus has been with limited etiologies. This study examined psychosocial functioning using the Behavioral Assessment System for Children, Third Edition (BASC-3) and the Hydrocephalus Outcome Questionnaire (HOQ) in children aged 5-17 years old. BASC-3 and HOQ parent report scores were compared between hydrocephalus etiologies. Medical factors (number of CSF diversion procedures, history of seizures, and years with hydrocephalus) and SES factors (family income, parent education, and parent occupational status) were examined as potential predictors for psychosocial outcomes. BASC-3 attention and executive functioning and HOQ social-emotional scales differed between etiologies. Years with hydrocephalus and a history of seizures were significant predictors for some BASC-3 scales and the HOQ social-emotional scale. SES variables did not predict any psychosocial outcomes examined. These results provide evidence that children who have had surgery for their hydrocephalus may be at increased risk of psychosocial difficulties, and that etiology and medical history may be contributing factors.

Keywords: hydrocephalus, BASC-3, neurosurgery, psychosocial

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Introduction

Hydrocephalus affects approximately one in 1000 births worldwide (Tamber, 2021). It generates a substantial emotional and financial burden for individuals, families, and health care centers. It is a disease characterized by obstruction or hypersecretion of cerebrospinal fluid resulting in ventriculomegaly, which causes disrupted white matter tract development and myelination, vascular damage, and chronic inflammation (Kahle et al., 2016; Tully & Dobyns, 2014). The sequelae from these processes are varied and complex depending on the etiology and severity of hydrocephalus (Kahle et al., 2016). Children with hydrocephalus often have additional medical conditions that impact development and prognosis. Due to the numerous issues associated with hydrocephalus, it is challenging to obtain a comprehensive picture of the child's health status and generate a holistic treatment plan. A better understanding of risk factors associated with health outcomes in hydrocephalus will improve management of this heterogenous condition.

Health Status in Hydrocephalus

In order to study health outcomes in pediatric hydrocephalus, it is helpful to consider how to define "health." There are many measures for evaluating health status and health-related quality of life (HRQoL) in pediatric neurosurgery. These measures evaluate different domains such as physical health, social functioning, impact of treatment, mental health, sleep disturbance, and caregiver concerns. Measures may be general, intended for use in any medical population, or they may be condition-specific. A recent review of HRQoL measures in pediatric neurosurgery examined 22 general and 25 condition-specific measures that are used in pediatric neurosurgery

research (Desai et al., 2019). Each of these measures varies in terms of what health domains they measure and what types of health outcomes are emphasized. General HRQoL measures are useful for capturing the overall functioning of patients and to track changes in health status over time. While general HRQoL measures such as the Health Utilities Index (HUI) (Furlong et al., 2001) capture a broad picture of health status, condition-specific measures allow clinicians to obtain information that may be unique or of higher importance to a given population. The Hydrocephalus Outcome Questionnaire (HOQ) is a well-validated 51-item hydrocephalus specific HRQoL questionnaire that provides a framework for measuring health status in pediatric hydrocephalus. The HOQ measures health status in three domains: cognitive, social-emotional (here used interchangeably with "psychosocial"), and physical health (Kulkarni, Rabin, et al., 2004). Given the potential benefits of using a condition-specific measure, the current study used the HOQ and its framework of three major health domains: physical, cognitive, and psychosocial. We specifically focused on the psychosocial domain of the HOQ, as this is an outcome category that is less studied in pediatric neurosurgery.

Children treated for hydrocephalus are routinely evaluated for functioning in the physical domain, such as sleep, eating habits, and mobility. Complications directly related to the hydrocephalus treatment, such as infection, shunt failure, or increasing ventricle size, are also closely monitored (Erps et al., 2018; Kahle et al., 2016; Kestle et al., 2016; Riva-Cambrin et al., 2016). Increasingly, cognitive function is a priority when evaluating neurosurgical outcomes. In particular, children with surgically-treated hydrocephalus generally have lower performance on standardized measures of intellectual functioning and more attention difficulties observed on both parent-report and standardized tests of attention (Arrington et al., 2016; Ball et al., 2013; Kulesz et al., 2015; Kulkarni et al., 2011; Swartwout et al., 2008). These deficits often persist

into adulthood (Jenkinson et al., 2011). Degree of cognitive impairment has also been found to be dependent on a number of factors, including etiology, medical comorbidities and surgical complications, race, family income, and other factors related to socioeconomic status (Karmur & Kulkarni, 2018; Kulkarni et al., 2008b; Swartwout et al., 2010). Less understood are the ways that psychosocial functioning is impacted in pediatric hydrocephalus. Despite some evidence that children with hydrocephalus have more problems with behavior and psychosocial functioning than their peers (Helder et al., 2011; Lindquist et al., 2006; Sumpter et al., 2012; Zielińska et al., 2017), this domain is often overlooked as a treatment consideration and a focus of research in the neurosurgery setting. To optimize treatment outcomes, all domains of health should be 1) well-characterized in this population in order to provide outcome expectations and recommendations to families and treatment teams and 2) routinely monitored in the neurosurgery setting as this is the primary medical point of contact for many children with hydrocephalus. This study used a general psychosocial measure, the Behavioral Assessment System for Children (BASC-3), in addition to the HOQ, in order to characterize psychosocial health in a pediatric hydrocephalus population.

Psychosocial Health in Hydrocephalus

Children with hydrocephalus appear to experience both internalizing and externalizing emotional symptoms at higher rates than the general population (Donders et al., 1992; Fletcher et al., 1995) and may have higher rates of autism (Lindquist et al., 2006) but there are few recent studies of psychosocial health in children with hydrocephalus. In a small study examining psychosocial functioning with the BASC-3, Helder and colleagues (2011) found that children with hydrocephalus have higher levels of depression, somatization, attention problems, and atypicality, but this study was limited to only one etiology: post-intraventricular hemorrhage

(bleeding in the ventricles of the brain). Additionally, the analysis did not correct for multiple comparisons, so the study may have over-represented psychosocial difficulties in these children. A more recent study using the Patient-Reported Outcomes Measurement Information System (PROMIS) scales reported no difference in self-reported levels of anxiety and depression in pediatric hydrocephalus patients compared to the normative population (Zimmerman, May, Barnes, Arynchyna, Alford, Wessinger, et al., 2020). The PROMIS was funded by the National Institutes of Health to generate health measures that are not disease-specific. There are individual PROMIS scales for various health concerns, such as sleep, pain, social functioning, anxiety, and depression (Cella et al., 2019). This study on self-reported levels of anxiety and depression was restricted to children who were able to complete a self-report questionnaire regarding their emotional state. The same research group separately reported self- and parent-report data using the Hydrocephalus Outcome Questionnaire. They found that children consistently rated their own health as better than did their parents (Zimmerman, May, Barnes, Arynchyna, Alford, Chagoya, et al., 2020), which may partially account for the finding of no elevated anxiety or depression in this pediatric hydrocephalus population. Additionally, the questions from the PROMIS scales are generally face valid with questions such as “I felt worried”. This may result in underreporting from children who tend to somaticize their emotional symptoms. There is evidence of differences in child self-reported quality of life relative to parent-report in a variety of conditions; however, the evidence is equivocal between studies (Kulkarni et al., 2008a; Lee et al., 2016). One study comparing parent- and self-report measures of anxiety found that for younger children, scores between parent- and self-report were negatively associated but for older children scores were positively associated (Shain et al., 2020). Children with ADHD have been found to report their quality of life as higher than their parents (Danckaerts et al., 2010) but a

recent meta-analysis did not find overall differences in parent- and self-report in children with ADHD (Lee et al., 2016). Adolescents with Autism Spectrum Disorder and typically-developing controls were found to have lower self-report symptoms of depression than their parents (Schwartzman & Corbett, 2020).

Given the variable nature of factors than can impact differences between parent- and self-report, the current study used parent-report of social and emotional functioning 1) in order to collect data for a larger patient population, including children who may not be able to complete self-report and 2) to remain consistent with the original design of the HOQ to be used as parent-report due to the potential for some children to be unable to complete self-report, either due to cognitive limitations or young age (Kulkarni, Rabin, et al., 2004).

Current research examining non-surgical hydrocephalus outcomes other than overall cognitive functioning typically addresses attention and executive function (Burmeister et al., 2005; Helder et al., 2011; Lacy et al., 2012; Mahone et al., 2002; Zabel et al., 2011). Attention processes have been primarily studied in spina bifida myelomeningocele (SBM). Greater deficits have been observed in posterior attention networks, proposed to regulate attention switching, as compared to anterior attention networks, proposed to regulate sustained attention. This is presumably due to greater disruption of white matter deficits in the posterior brain in SBM due to the presence of Chiari II malformation (Copp et al., 2015). Executive function generally refers to one's ability to reason, problem solve, and plan (Blair, 2017). Executive functioning is a component of psychosocial and behavioral functioning as it can impact a child's ability to regulate and express their emotions and interact with others (Yeates et al., 2007). The ability to plan and reason strongly influences behavior and social functioning, but measures of executive functioning do not capture internalizing symptoms such as depression and anxiety as well as they

capture externalizing symptoms such as hyperactivity and impulsivity. Examining individual internalizing and externalizing symptoms such as anxiety, depression, hyperactivity, and inattention in addition to executive functioning may help to target intervention for children with hydrocephalus who have psychosocial problems. For the purposes of this study, executive functioning was examined as an aspect of psychosocial and behavioral functioning.

Hydrocephalus Etiologies

Clinical course and prognosis for children with hydrocephalus can vary widely according to the cause of the hydrocephalus. Due to the variability in clinical course, research conducted on non-surgical outcomes in pediatric hydrocephalus is often limited to one or two etiologies (Helder et al., 2011; Jenkinson et al., 2011; Kulesz et al., 2015; Lindquist et al., 2006; Swartwout et al., 2008; Zabel et al., 2011) or does not directly compare etiologies (Donders et al., 1992; Fletcher et al., 1995; Sumpter et al., 2012; Zimmerman, May, Barnes, Arynchyna, Alford, Chagoya, et al., 2020; Zimmerman, May, Barnes, Arynchyna, Alford, Wessinger, et al., 2020).

The present study included the following hydrocephalus etiologies: post-IVH secondary to prematurity, myelomeningocele, communicating congenital hydrocephalus, aqueductal stenosis (AS), and other, as diagnosed by the neurosurgeons managing hydrocephalus interventions. These etiologies (post-IVH secondary to prematurity, myelomeningocele, communicating congenital hydrocephalus, aqueductal stenosis) are the four largest etiology groups in the current study population. They are also of interest due to indications that there are differences between these etiologies in terms of health outcomes.

Post-IVH Secondary to Prematurity

Post-IVH secondary to prematurity refers to children who develop hydrocephalus after bleeding in the ventricles of their brain occurs associated with premature birth. HRQoL in

children with hydrocephalus due to intraventricular hemorrhage (IVH) due to premature birth is generally worse than that of typically developing children (Gigi et al., 2019; Robinson, 2012) but there are few studies comparing post-IVH hydrocephalus to other etiologies. In one study, premature birth and hydrocephalus etiology (myelomeningocele, IVH due to premature birth, aqueductal stenosis, postinfectious, congenital communicating, posterior fossa cyst, and other) were found to be associated with lower physical health HOQ scores, but not cognitive or social-emotional HOQ scores (Kulkarni & Shams, 2007). Another research group found lower HRQoL in motor, emotional, social, and school/daycare domains of functioning for children with shunted hydrocephalus due to prematurity compared to infants born premature with normal neonatal cranial ultrasound studies (Gigi et al., 2019). This indicates that the presence of hydrocephalus and its associated complications may be partially responsible for lower HRQoL above and beyond the impact of premature birth. They also found that worse personal-social functioning as measured by the Battelle Developmental Inventory, a screener of developmental milestones, was associated with lower overall HRQoL. This highlights psychosocial functioning as an important domain for examining developmental outcomes in hydrocephalus due to IVH secondary to premature birth.

Myelomeningocele

Myelomeningocele is the most severe form of spina bifida and is characterized by a protrusion of the spinal cord through the spinal vertebrae. This protrusion must be surgically closed, and children with spina bifida myelomeningocele (SBM) often have a number of secondary effects, including Chiari II malformation, genitourinary dysfunction, sensory and motor problems, and hydrocephalus (McCarthy et al., 2019). In children with myelomeningocele, comorbid conditions can vary between patients according to the location of

the myelomeningocele. Children with upper level (T-12 and above) spinal lesions were found to have lower scores on measures of intelligence, academic functioning, and adaptive skills. Upper level lesions were also associated with lower cerebrum and cerebellar volumes, though they were not associated with qualitative cerebellar abnormalities (Fletcher et al., 2005). Depending on the level of spinal lesion, children with SBM may have comorbid neurogenic bowel and bladder and severe mobility issues. This is a potential source of variability when measuring psychosocial outcomes, as this may impact social functioning and ratings of quality of life. It is important to keep in mind, however, that patients with spinal cord lesions may still have high quality of life, and there is evidence of patient vs. physician disparities in perceived quality of life (Cushman et al., 2015). Additionally, degree of psychosocial impairment may be mediated by factors in addition to level of spinal lesion, such as participation in and satisfaction with activities (Müller et al., 2017). .

Spina bifida myelomeningocele is associated with Chiari II malformation in about 90% of cases (Copp et al., 2015). Chiari II involves herniation of the cerebellum and reorganization of the cerebellum with cerebellar volume reductions. It has been associated with reduced hippocampal volume and about one third to one half of children with Chiari II malformation have hypogenesis of the corpus callosum. White matter tracts connecting anterior to posterior brain regions are reduced, with posterior white matter generally more impacted. As a result, children with Chiari II malformation and hydrocephalus often have deficits in cognitive tasks requiring posterior attention networks (e.g., orienting or switching attention). Degrees of deficits are associated with degree of white matter abnormalities (Copp et al., 2015).

Clearly, there is evidence of variability within hydrocephalus etiologies. As a result, there are few studies comparing SBM-associated hydrocephalus to other etiologies relative to studies

examining SBM alone. Of those examining multiple etiologies, studies comparing myelomeningocele and AS have uncovered differences in executive function, sustained attention, and memory, with the myelomeningocele group typically demonstrating greater deficits. (Hampton et al., 2013; Swartwout et al., 2008). Interestingly, presence or absence of cerebellar abnormalities differentiated the aqueductal stenosis group in terms of cognitive functioning; children with AS with cerebellar abnormalities performed worse than those without. (Hampton et al., 2013). The impact of cerebellar involvement in SBM may explain why AS patients with cerebellar abnormalities look more similar to SBM patients than AS patients without cerebellar abnormalities.

While the present study did not have an adequate sample size to separate myelomeningocele patients by spinal lesion level or specific white matter abnormalities, comparing SBM-associated hydrocephalus to other etiologies was considered an important first step in understanding the extent of variability in psychosocial outcomes within and between etiologies.

Communicating Congenital Hydrocephalus

Communicating congenital hydrocephalus refers to any cause of hydrocephalus that was present at birth where the cerebrospinal fluid can still move between the ventricles but is blocked after exiting the ventricles. There may also be abnormalities in the re-absorption of CSF into the ventricles. There has been some debate about how to precisely define communicating hydrocephalus due to the acknowledgment that some etiologies classified as “communicating” can include some obstruction in the ventricular system (Agarwal et al., 2016). For the purposes of this study, neurosurgeons at the study site classified an etiology of “communicating congenital hydrocephalus” for cases where CSF appeared to be able to move through the ventricles without

obstruction.

Aqueductal Stenosis

Aqueductal stenosis (AS) is the most common cause of congenital hydrocephalus and is characterized by obstruction between the third and fourth ventricle (Kahle et al., 2016). One study that compared multiple hydrocephalus etiologies (myelomeningocele, IVH due to premature birth, aqueductal stenosis, post-infectious, congenital communicating hydrocephalus, posterior fossa cyst, and other) found differences in physical health status but not in social-emotional health as measured by the HOQ (Kulkarni & Shams, 2007). They reported that physical health was better in children with an etiology of IVH due to premature birth or aqueductal stenosis relative to other etiologies. Intellectual functioning in children with AS has been reported to be both higher than and lower than that of children with communicating congenital hydrocephalus in a review of older studies (1980's-1990's) of hydrocephalus outcomes (Cinalli et al., 2011)

Medical Factors Impacting Health Outcomes

Number of CSF Diversion Surgeries

In addition to etiology, a number of other factors can impact health status in children with hydrocephalus. A history of shunt revisions (repeated surgery after a shunt is placed due to shunt malfunction) is a risk factor for future shunt infections (Erps et al., 2018). Number of shunt revisions has been associated with worse cognitive functioning in children (Arrington et al., 2016), however, in adults who previously had pediatric hydrocephalus, shunt revision history was not associated with cognitive function, quality of life, or disability (Jenkinson et al., 2011; Kutscher et al., 2015). One study found that number of neurosurgical procedures was associated with worse physical health scores on the HOQ (Ros-Sanjuán et al., 2021) but not with cognitive

or social-emotional health. However, this study was limited to patients who had an endoscopic third ventriculostomy (ETV), (a type of neurosurgical intervention that does not involve a shunt) and the researchers used a Spanish version of the HOQ. It is unclear how cultural and language factors may impact parent report of their child's health status, making it difficult to know if these results would generalize to an English-speaking population. Additionally, the majority of permanent CSF diversion surgeries in pediatric hydrocephalus are shunts (Pindrik et al., 2020), making this an important neurosurgical procedure to include when examining outcomes. In a study of prematurely born infants with and without hydrocephalus, number of neurosurgical procedures was not associated with HRQoL, though the total sample size for the study was small (n = 40) and the procedures were not limited to CSF diversion surgeries (Gigi et al., 2019). In order to better understand how number of CSF diversion surgeries may impact psychosocial functioning in a pediatric hydrocephalus population, the present study included total number of initial shunt placements and shunt revisions, ETVs and ETV revisions, new shunt placements, and extra-ventricular drain (EVD) placements.

Duration Since First Permanent CSF Diversion Surgery

Age at first neurosurgery for hydrocephalus has been identified as a risk factors for future shunt infection, which can lead to additional medical complications (Simon et al., 2012). Age at first surgery has been examined as a potential factor for cognitive, social-emotional, and physical health using the HOQ, with some studies showing it did not have a significant impact on outcomes (Karmur & Kulkarni, 2018; Kulkarni et al., 2008b) and other indicating it may have an impact on cognitive functioning as measured by the HOQ (Kulkarni & Shams, 2007). Reynolds and colleagues (Reynolds et al., 2021) examined number of years with hydrocephalus, measured by time since first hydrocephalus surgery in relation to overall HOQ score and parental concern

scores. They found years with hydrocephalus was associated with parental concern but not overall HOQ score. In the present population, the majority of patients received their first permanent CSF diversion surgery in the first year of life (67%). Therefore, rather than examining age at first surgery, we examined years with hydrocephalus as a factor in psychosocial health outcomes. This allows examination of the impact of hydrocephalus on psychosocial functioning through development and lays the groundwork for future research examining long vs. short-term outcomes for this population.

Seizures

Seizure history is another medical factor that has been identified as a significant risk factor for poorer HRQoL in pediatric hydrocephalus (Gigi et al., 2019; Kulkarni et al., 2008b; Kulkarni & Shams, 2007). Kulkarni and colleagues reported that increased seizure frequency in a mixed-etiology sample was associated with worse HOQ scores in all three domains of health-related quality of life: physical, cognitive, and social-emotional (Kulkarni & Shams, 2007). Gigi and colleagues reported the presence of any neonatal seizures in a post-hemorrhagic hydrocephalus population was associated with worse HRQoL as measured by the Pediatric Quality of Life Inventory (PedsQL) (Gigi et al., 2019). In the present study, any history of any seizures was examined in order to determine if this medical risk factor contributes to psychosocial difficulties in our sample.

Non-Medical Factors Impacting Health Outcomes

It is well-established that socioeconomic status (SES) can impact health outcomes in children (Poulain et al., 2020; Reiss, 2013). SES has been fairly well-studied in pediatric hydrocephalus, however, variables used as proxies for SES are often less than optimal. For example, type of insurance (private vs. public) is often used as a proxy for SES (Attenello et al.,

2015; Reynolds et al., 2020). A single variable as an estimate of SES may overlook important information regarding how social and environmental variables can impact health outcomes. Of the studies that collected additional SES variables, such as family income and parent occupational status, SES has been shown to have significant effects on the degree of impact of hydrocephalus on the family (Agajany et al., 2019), verbal abilities (Swartwout et al., 2010), and overall and physical health (Karmur & Kulkarni, 2018). Limited research has been done on the impact of SES specifically on psychosocial and behavioral functioning in children with hydrocephalus. The present study examined family income, parent education, and parent occupational status as important aspects of SES in order to obtain more detailed information on how these non-surgical factors can impact psychosocial outcomes in pediatric hydrocephalus.

The current study addressed several gaps in the pediatric hydrocephalus literature. Despite several older studies specifically examining social, emotional, and behavioral functioning in pediatric hydrocephalus, there are few recent published studies. It is important to re-examine this domain of functioning in this population as medical management for hydrocephalus has changed significantly in the past 20 years. Increased use of non-shunting hydrocephalus management interventions such as ETV as well as advances in imaging and monitoring of hydrocephalus may result in different trajectories for patients. Additionally, due to the medical complexity of many pediatric hydrocephalus patients, psychosocial concerns may be overlooked when medical management must take priority. The HOQ is a promising measure for use as a screener for psychosocial difficulties in a neurosurgery setting. Continued research with this measure alongside increased understanding of the psychosocial functioning of this population will allow for better integrated care in pediatric hydrocephalus.

Aims and Hypotheses of the Current Study

The primary purpose of this study was to examine psychosocial functioning in children who have undergone surgical treatment for hydrocephalus of various etiologies. The secondary purpose of the study was to identify potential medical and non-medical risk factors that impact psychosocial health in this population.

Aim 1

The first aim was to examine psychosocial functioning in a pediatric hydrocephalus population using an established measure of behavioral and psychosocial functioning, the Behavioral Assessment System for Children (BASC-3), and a hydrocephalus-specific questionnaire, the HOQ. Patients were grouped according to etiology in order to identify differences between groups in terms of psychosocial functioning.

Hypothesis 1. I hypothesized that etiology would be significantly associated with psychosocial functioning. Since myelomeningocele has previously been demonstrated to be a risk factor for worse overall health (Kulkarni et al., 2008b) and neuropsychological functioning (Hampton et al., 2013; Swartwout et al., 2008), I expected that children with an etiology of myelomeningocele would have worse BASC-3 scores as well as worse HOQ social-emotional scores compared to other etiologies. Due to limited existing data on health outcomes in other hydrocephalus etiologies, no directional hypothesis was presented for how other etiologies such as aqueductal stenosis would rank in terms of BASC-3 and HOQ scores.

Aim 2

The second aim was to examine surgical and medical risk factors for worse psychosocial functioning. History of seizures, years since first permanent CSF diversion surgery, and total

number of CSF diversion surgeries were examined for their relative contributions to psychosocial health.

Hypothesis 2. I hypothesized that a history of seizures, more years with hydrocephalus, and a higher number of surgeries would be associated with worse scores on the BASC-3 and the HOQ social-emotional health scales.

Aim 3

The third aim of this study was to examine SES related risk factors for worse psychosocial functioning. Parent education, income, and employment status were examined for their relative contributions to psychosocial health.

Hypothesis 3. I hypothesized that higher family income, higher parental education, and having at least one parent with full time employment status would be associated with better BASC-3 and HOQ scores.

Methods

Participants

Approval for this study was obtained from the Institutional Review Board at the University of Utah and Brigham Young University. We recruited caregivers of children 5-17 years old with hydrocephalus to assess their physical, social-emotional, and cognitive functioning using the HOQ and their behavioral and psychosocial functioning using the BASC-3. Participants were identified from the Hydrocephalus Clinical Research Network (HCRN) core data project (i.e., Registry). All patients who receive surgical intervention for hydrocephalus at Primary Children's Hospital are entered in the Registry. There were 397 eligible patients in the Registry. Sixty-six families had already completed both surveys as a part of a pilot study verifying the psychometric properties of the HOQ's social-emotional domain (Wall et al., 2021). Twenty-three caregivers

had previously declined to participate. The remaining potentially eligible participants were called for potential participation in the present study (see Table 1 for participant and non-participant characteristics). One hundred twenty-five participants completed both surveys. Average age was 9.8 years, with a range from 5.2 to 17.9 and 38% of patients were female. This is similar to the demographics of the Registry population (Table 1). Distribution of etiologies across the sample and total number of CSF diversion surgeries were similar to that in the Registry. Age at first permanent CSF diversion surgery was slightly higher for the sample compared to the Registry population (914 days vs. 731 days) but this was not statistically significant, ($t = 1.29$, $p = .20$). Race and ethnicity are not collected in the Registry, so it is unknown if there were differences between the sample and the Registry population.

Parents/caregivers of all patients with an etiology entered in the Registry were eligible for participation, with the exception of those meeting the exclusion criteria listed below. Registry patients without a recorded etiology or with a patient status form indicating the child has died or the family has moved out of network were excluded. The Hydrocephalus Outcome Questionnaire (HOQ) was developed for use with children 5 years of age and older, so those under 5 years old and older than 17 (the age at which many children transfer from pediatric to adult care) were excluded. Although the HOQ has been translated and used in Spanish (Iglesias et. al., 2018) validity and reliability evidence for the Spanish version is limited, therefore, only the English version was used. Families who are unable to complete the English versions of the HOQ and the Behavior Assessment System for Children (BASC-3) were excluded.

Screening and Enrollment

I reviewed the HCRN Registry to generate a list of potentially eligible participants. The parents/guardians of eligible patients were sent a letter to discuss the study. Eligible participants

were contacted via telephone 1-3 weeks after the letter was sent to see if they would like to participate. All surveys were completed online. The Registry's unique subject identifiers were used for all study participants to maintain participant confidentiality.

Data Collection

The HOQ is a 51-item questionnaire with questions about physical, cognitive, and social-emotional functioning (Kulkarni, Rabin, et al., 2004). A link to the questionnaire was emailed to families to complete the questionnaire online via a REDCap survey. The first page of the REDCap survey was the consent document. Background information not found in the Registry (history of seizures and SES variables) were asked on the second page of the REDCap survey. The HOQ began on page three of the REDCap survey.

The BASC-3 is a 139 to 175 item questionnaire (depending on age range) examining behavioral and emotional functioning in children (Reynolds & Kamphaus, 2015). A link to the questionnaire was emailed to participating families.

Data from the Registry was extracted and included etiology of hydrocephalus, age at time of first surgery, and number and type of neurosurgical procedures. Neurosurgical procedures included were primary shunt placements, shunt revisions, extraventricular drains (EVDs) and endoscopic third-ventriculostomies (ETVs).

All completed questionnaires were reviewed for missing data. BASC-3 scales can be generated if limited data is missing. If too many items were missing to be able to generate a BASC-3 scale, the sample size for a given analysis was adjusted to reflect the number of BASC-3 scales able to be generated. HOQ scores can be generated with missing data by adjusting the total points relative to the number of questions answered.

Data Storage

Registry data is managed by the Data Coordinating Center at the University of Utah. The following is the description of information security included in all DCC studies:

The DCC is housed in a building with 24-hour on-site security guards. The server facility (a dedicated, locked 720 ft^2 server room) is locked separately from the remainder of the DCC and access to the building is monitored by security personnel year-round. The DCC coordinates its network infrastructure and security with the Health Sciences Center (HSC) information systems at the University of Utah. This provides the DCC with effective firewall hardware, automatic network intrusion detection, and the expertise of dedicated security experts working at the University. Network equipment includes four high-speed switches. User authentication is centralized with three Windows 2012 domain servers. Communication over public networks is encrypted with virtual point-to-point sessions using Transport Layer Security (TLS) or virtual private network (VPN) technologies, both of which provide at least 128-bit encryption. Our web-based clinical studies data management system, and eRoom™ (Web-based collaborative workspace) and other web applications use the TLS protocol to transmit data securely over the Internet. Direct access to DCC machines is only available while physically located inside the DCC offices, or via a VPN client. All network traffic is monitored for intrusion attempts, security scans are regularly run against our servers, and our IT staff are notified of intrusion alerts.

Security is maintained with Windows 2012 user/group domain-level security. Users are required to change their passwords every 90 days, and workstations time out after 5 minutes of inactivity. All files are protected at group and user levels; database security is handled in a similar manner with group-level access to databases, tables, and views in Microsoft SQL Server.

REDCap and Pearson's web-based assessment system do not require individually identifiable information to be collected and are HIPAA compliant. Participants were identified using the unique numerical identifier already assigned to them in the Registry. Data extracted from the questionnaires and the Registry were recorded using participant's numerical identifiers. Once consented, only necessary potentially identifying information was retained in the study files (i.e., date of birth and dates of surgical treatment). These were required to conduct necessary analyses for the study such as years with hydrocephalus.

Measures

Behavioral Assessment System for Children, Third Edition (BASC-3)

The proposed study used the BASC-3 Parent Rating Scales (PRS), a commonly used measure for assessing behavioral and emotional problems in children that has undergone extensive reliability and validity testing (Reynolds & Kamphaus, 2015). There are 3 PRS versions; one for preschool age children, ages 2-5, one for children ages 6-11, and one for adolescents, ages 12-21. The norms were obtained from a large, representative sample of children across the United States. Items for the PRS are written at a 4th grade reading level, making the measure accessible to most parents, regardless of education level.

Clinical scales for the BASC-3 were developed by comparing responses between children with no identified clinical diagnosis and those with general clinical problems, ADHD, autism, or emotional or behavioral disturbance. The following clinical scales were generated, which reliably differentiate between typical children and those with clinical presentation of emotional or behavioral problems: aggression, anxiety, attention problems, atypicality, conduct problems, depression, hyperactivity, learning problems, somatization, and withdrawal. In addition, the adaptive scales (activities of daily living, adaptability, functional communication, leadership,

social skills, study skills) were developed to assess for functioning in a number of positive domains. Some scales (e.g., conduct and leadership) are not generated for the preschool age range. Content scales (anger control, bullying, developmental social disorders, emotional self-control, executive functioning, negative emotionality, resiliency), composite scales (externalizing problems, internalizing problems, adaptive skills, behavioral symptoms index), a clinical probability scale, and a functional impairment scale were developed as well. The present study qualitatively examined all clinical scales according to etiology, however, only select scales were used in analyses. The anxiety, depression, attention problems, hyperactivity, and executive functioning scales were used in analyses in order to examine select components of psychosocial functioning that are hypothesized to have a greater impact on overall quality of life (Danckaerts et al., 2010; Fletcher et al., 1995); as well as to balance the utility of looking at multiple scales with the increased risk of Type I error with multiple comparisons. For all scales, scores are reported as T-scores with a mean of 50 and SD of 10. A T-score of 60-69 is considered "at risk" and a T-score of 70 or above is considered "clinically significant." The BASC-3 also includes validity indexes, such as the F-index, for "faking bad", the L-index for "faking good", and the response pattern index to identify inattentiveness to items. Items are answered with a 4 level Likert scale (never, sometimes, often, always). The BASC-3 contains items like "my child is sad" and "my child is nervous."

Internal consistency across ages and gender ranged from .91-.97 for the internalizing problems composite scale, .88-.97 for the externalizing problems composite scale, and .88-.93 for the executive functioning content scale in the general norm samples. Test-retest reliability was measured by repeat administration to the same caregiver 7-70 days after the first administration. Correlations for the internalizing and externalizing composite and executive

function content scales ranged from r = .87 to r = .93. Inter-rater reliability was assessed by having two different caregivers fill out the PRS, all administered within 0-63 days of each other. Correlations for the internalizing and externalizing composite and executive function content scales ranged from r =.59 to r = .86. Some variability is expected between different caregivers as each is likely to observe different components of the child's behavior. Moderate to high correlation between the BASC-3 with the Achenbach System of Empirically Based Assessment Child Behavior Checklist (ASEBA CBCL), Conners 3, Autism Spectrum Rating Scale (ASRS), and Delis Rating of Executive Function indicate good construct validity in that the BASC-3 is adequately measuring problems with behavioral and emotional functioning.

Hydrocephalus Outcome Questionnaire

The Hydrocephalus Outcome Questionnaire (HOQ) is a 51-item instrument designed to measure the physical, cognitive, and social-emotional functioning of children age 5-17 years with hydrocephalus (Kulkarni, Drake, et al., 2004). The questionnaire is parent report and items are answered with a 5 item Likert scale (not at all true, a little true, somewhat true, quite a bit true, very true). The HOQ provides scores from 0.0 (worse health status) to 1.0 (better health status). Each item on the 5-point scale receives a score of 0 (worse health status) to 4 (better health status). Scores of items are summed then divided by the highest possible summed score to provide a final score of 0 to 1. The social-emotional domain contains items like "My child is often irritable" or "My child worries about the future." The HOQ was developed in 2004 at the Hospital for Sick Children, Toronto, with a population of 90 children in whom hydrocephalus was diagnosed at least 6 months prior to administration of the measure and who were at least 5 years old. The HOQ has since been used in a number of studies examining health outcomes in children with hydrocephalus.

Internal consistency from the initial test development, measured with Cronbach α, was 0.94 for overall health, 0.93 for physical health, 0.82 for social-emotional health, and 0.91 for cognitive health. Test-retest reliability, with the questionnaire re-administered a mean of 18 days apart, was 0.93 for overall health, 0.98 for physical health, 0.84 for social-emotional health, and 0.92 for cognitive health. Responses from mother-father pairs who both filled out the HOQ were evaluated for interrater reliability, with a correlation of 0.88 for overall health, 0.96 for physical health, 0.76 for social-emotional health, and 0.76 for cognitive health.

Content validity was evaluated by an expert panel of pediatric neurosurgeons to ensure adequate breadth of coverage of health status items for children with hydrocephalus. Construct validity was evaluated by correlating HOQ scores with several established measures of health, the WeeFIM, a measure of physical and overall health, the SDQ, a measure of social-emotional health, the WRAT, a measure of cognitive health, and the HUI-2, a measure of overall health. Correlations were moderate to high (.59-.89) between HOQ domains and their corresponding external measure. Since its development, the HOQ has been compared to a comprehensive neuropsychological battery (Kulkarni et al., 2011) and found to have good convergent validity between the cognitive domain and neuropsychological tests, as well as divergent validity between the physical and social-emotional domains and neuropsychological tests. The present study will provide additional evidence of the characteristics of children as measured by the HOQ in relation to health risk factors.

Data Analysis

The primary purpose of the proposed study was to examine the relationship between etiology and psychosocial health outcomes as measured by the BASC-3 and HOQ. A total of 125 participants completed both surveys. Prior to performing analyses, data were cleaned and

prepared. All continuous variables for which means would be compared were examined for outliers. Three depression and four anxiety composite scores appeared to be outliers (e.g., beyond the median value +/- 2 interquartile ranges). The BASC-3 reports for these outliers were examined individually, and the response patterns appeared to be an accurate representation of the functioning of those children based on the responses for individual questions included in validity scales, so the data points were left unchanged and no transformations were conducted on the data. Two BASC reports had too many missing items to be able to generate scales and were therefore excluded from BASC-3 analyses. One additional BASC-3 report was missing the executive function, leaving n = 122 for BASC-3 executive functioning analyses, and n = 123 for the remaining BASC-3 analyses. There were five subjects with some missing social-emotional HOQ items. This generally occurs if there is an item a caregiver feels does not apply to their child. The maximum number of missing items for a participant was 5 (out of 24 possible items). Social-emotional domain score totals and percentages were adjusted for these subjects.

The Kruskal-Wallis test for one continuous variable and one categorical variable was used to examine differences between etiology of: post-IVH secondary to prematurity, myelomeningocele, communicating congenital hydrocephalus, aqueductal stenosis, and other in terms of BASC-3 anxiety, depression, hyperactivity, attention problems, and executive function scales and HOQ social-emotional scores. For significant Kruskal-Wallis tests, Dunn's test was used to examine which etiologies groups differed. For the BASC-3, general combined age adjusted T-scores were generated by the web-based scoring platform.

Linear regression was used for examining relationships between surgical and medical risk factors and health outcomes. Specifically, a separate regression analysis with number of neurosurgical procedures (nsproc, continuous), presence of seizures (seiz, categorical), and years

with hydrocephalus (years, continuous) as the predictors were completed for each of the following outcomes: HOQ social-emotional score, BASC-3 anxiety clinical scale, BASC-3 depression clinical scale, BASC-3 attention problems clinical scale, BASC-3 hyperactivity clinical scale, and BASC-3 executive function content scale. History of seizures was dummy coded (yes, no). R^2 was used to examine the proportion of variance in the outcome explained by the predictors. Regression models used are as follows:

$HOQSE_i = b_0 + b_1nsproc_i + b_2seiz_i + b_3years_i$

$Anx_i = b_0 + b_1nsproc_i + b_2seiz_i + b_3years_i$

$Dep_i = b_0 + b_1nsproc_i + b_2seiz_i + b_3years_i$

$Attn_i = b_0 + b_1nsproc_i + b_2seiz_i + b_3years_i$

$Hyper_i = b_0 + b_1nsproc_i + b_2seiz_i + b_3years_i$

$ExFx_i = b_0 + b_1nsproc_i + b_2seiz_i + b_3years_i$

Linear regression was similarly used to examine relationships between SES variables and health outcomes. A separate regression analysis with parent education (educ, categorical), family income (income, categorical), parent job status (empl, categorical) was completed for each of the following outcomes: HOQ social-emotional score, BASC-3 anxiety clinical scale, BASC-3 depression clinical scale, BASC-3 attention problems clinical scale, BASC-3 hyperactivity clinical scale, and BASC-3 executive function content scale. The following dummy codes were used: parent education (some high school, completed high school, completed 2-year degree, completed 4-year degree, completed master's degree, completed doctorate degree), income ($0-$20,000, $20,000-$40,000, $40,000-$60,000, $60,000-$80,000, $80,000-$100,000, >$100,000), and employment (at least one parent full time employed, no parent full time employed). Regression models used are as follows:

$HOQSE_i = b_0 + b_1income_i + b_2educ_i + b_3empl_i$

$Anx_i = b_0 + b_1income_i + b_2educ_i + b_3empl_i$

$Dep_i = b_0 + b_1income_i + b_2educ_i + b_3empl_i$

$Attn_i = b_0 + b_1income_i + b_2educ_i + b_3empl_i$

$Hyper_i = b_0 + b_1income_i + b_2educ_i + b_3empl_i$

$ExFx_i = b_0 + b_1income_i + b_2educ_i + b_3empl_i$

The cutoff for significance was $\alpha = .05$. Given the relatively low number of regression analyses conducted and in order to compare the current study to prior studies using the HOQ, corrections for multiple comparisons were not used. Regressions were completed using the regress command and Kruskal Wallis and Dunn's test completed using the kwallis and dunntest commands in Stata.

Results

The mean social-emotional HOQ score was 0.70. HOQ physical health, cognitive health, and total HOQ averages were 0.63, 0.54, and 0.64, respectively (Table 8). The Kruskal-Wallis test examining the relationship between etiology and BASC-3 anxiety, depression, hyperactivity, attention problems, and executive function scales and HOQ social-emotional scores revealed a significant association between etiology and BASC-3 attention score, $\chi^2 = 10.28$, $p < .05$, BASC-3 executive functioning score, $\chi^2 = 10.55$, $p < .05$ and HOQ social-emotional score, $\chi^2 = 10.57$, $p < .05$, but not between etiology and BASC-3 anxiety, depression, or hyperactivity scores (Table 2). Post hoc analyses revealed a significant difference between etiology of IVH secondary to prematurity compared to myelomeningocele ($p = .01$), aqueductal stenosis ($p < .01$), and other ($p < .001$) for BASC-3 executive functioning scores. There was similarly a difference between etiology of IVH secondary to prematurity compared to myelomeningocele (p

< .01), aqueductal stenosis ($p < .01$), and other ($p < .01$) for BASC-3 attention scores. IVH secondary to prematurity differed from aqueductal stenosis ($p = .03$) and other ($p < .01$) on HOQ social-emotional scores. Myelomeningocele differed from other ($p = .02$) on HOQ social-emotional scores.

Linear regression analyses for medical factors (number of neurosurgical procedures, history of seizures, and years with hydrocephalus) revealed an overall significant model for BASC-3 anxiety, ($F(3, 119) = 3.30$, $p = .02$, $R^2 = .08$), attention, ($F(3, 119) = 5.36$, $p = .002$, $R^2 = .12$), hyperactivity, ($F(3, 119) = 3.38$, $p = .02$, $R^2 = .08$), and executive functioning scales, ($F(3, 118) = 7.61$, $p < .001$, $R^2 = .16$) (Table 3). The individual predictors were examined further and revealed that any history of seizures was associated with lower BASC-3 anxiety scores ($t = -2.10$, $b = -5.43$, 95% CI [-10.54, -0.31], $p = .04$) and lower (worse social-emotional health) HOQ social-emotional scores ($t = -2.26$, $b = -.09$, 95% CI [-.16, -.01], $p = .03$). Years with hydrocephalus was associated with higher BASC-3 attention ($t = 3.48$, $b = 1.06$, 95% CI [0.46, 1.66], $p = .001$), hyperactivity ($t = 1.96$, $b = 0.72$, 95% CI [-0.008, 1.45], $p = .05$), and executive functioning scores ($t = 3.86$, $b = 1.24$, 95% CI [0.60, 1.87], $p < .001$), and lower HOQ social-emotional scores ($t = -2.51$, $b = -.01$, 95% CI [-0.02, -.003], $p = .01$).

Linear regression analyses for SES factors (family income, parent education, and parent occupational status) did not indicate an overall significant model for any BASC-3 or HOQ scores (Table 4).

Discussion

The primary purpose of this study was to examine the relationship between etiology and psychosocial health outcomes as measured by the BASC-3 and HOQ and to identify medical and non-medical factors that may impact these outcomes.

Etiology and Psychosocial Functioning (Hypothesis 1)

The first aim of this study was to examine the relationship between hydrocephalus etiology (post-IVH secondary to prematurity, myelomeningocele, communicating congenital hydrocephalus, aqueductal stenosis, and other) and BASC-3 anxiety, depression, attention, hyperactivity, and executive functioning and HOQ social-emotional scores. I hypothesized that the myelomeningocele groups would have higher BASC-3 and lower HOQ scores (indicating worse functioning) than other etiology groups. There was a significant difference between etiology groups in BASC-3 attention and executive functioning. Attention and executive functioning scores were highest in the post-IVH secondary to prematurity group, with an average t-score of 63 and 66 respectively, followed by communicating congenital hydrocephalus, with an average t-score of 60 for attention and 61 for executive functioning. The average executive functioning and attention scores for these two groups were in the "at-risk" range (one to two standard deviations above the mean), compared to mean scores in the "average" range (within one standard deviation of the mean) for all other etiologies. HOQ social-emotional scores also differed between etiologies, with the post-IVH secondary to prematurity group having the lowest scores (corresponding with lower functioning), followed by myelomeningocele, then communicating congenital hydrocephalus. These results indicate that etiology is an important consideration when evaluating psychosocial outcomes, but in the present study population, myelomeningocele was not the etiology with the highest risk of psychosocial difficulties.

The present results revealed significant etiology group differences between group BASC-3 scores only in executive functioning and attention. Executive functioning and attention are common non-surgical factors studied in pediatric hydrocephalus (Burmeister et al., 2005; Helder et al., 2011; Lacy et al., 2012; Mahone et al., 2002; Zabel et al., 2011). Although children with

hydrocephalus are consistently found to have significant differences in executive functioning and attention on both standardized neuropsychological exams (Burmeister et al., 2005; Lacy et al., 2012) and parent-report measures (Helder et al., 2011; Mahone et al., 2002; Zabel et al., 2011) compared to healthy controls or normative populations, average parent-report scores are often not elevated to a level of clinical significance. It is possible that this is due to the use of single etiologies or combined etiologies in many of these studies. The present study revealed some etiologies with means 1-2 standard deviation range above average and other etiologies with means in the average range (Table 2). This indicates that it may be important for future research to clarify that some etiologies may be more likely than others to demonstrate these difficulties. In their proposed model of social competence in the context of pediatric brain injuries, Yeates and colleagues discuss that brain regions involved in cognitive and executive functioning abilities may overlap some with brain regions implicated in social-cognitive and emotional functioning, but not entirely (Yeates et al., 2007). They additionally discuss that regions of brain insults can result in differing psychosocial symptoms (e.g., a right frontal lesion is more associated with emotionality and disinhibition while a left frontal lesion is more associated with depression and withdrawal). This highlights how hydrocephalus etiology, and severity within an etiology group, may impact psychosocial functioning, depending on which brain regions have been impacted. Future research may elect to include all etiologies and examine results according to etiology due to the evidence from the current study that elevations in psychosocial difficulties may be obscured when etiologies are limited or all etiology scores are combined.

The present results provide information about what symptoms associated with psychosocial functioning may be more likely to present in different etiologies. Children with an etiology of post-IVH secondary to prematurity or communicating congenital hydrocephalus may

be more likely to have executive functioning and attention deficits, while children with hydrocephalus of any etiology may be equally likely to experience other psychosocial difficulties, such as depression or hyperactivity. Although average BASC-3 scores for the scales examined in the primary analyses for myelomeningocele, aqueductal stenosis, and other etiologies; and for all etiologies combined were in the "average" range (Table 2), the proportion of participants with scores in the "at-risk" or "clinically significant" range across scales was higher than would be expected in a normally distributed population (Table 5). Additionally, exploratory analyses comparing BASC-3 scores of all etiologies combined to the normative population mean (T-score = 50) with a Bonferroni's correction for multiple t-tests revealed a significant difference from normative population means for depression, attention, hyperactivity, and executive functioning, but not for anxiety (Table 6).

Variability in Psychosocial Functioning

As a whole, parents and caregivers of children with hydrocephalus seem to report higher levels of depression, attention, hyperactivity, and executive functioning difficulty compared to the normative population, but averages are generally not to a level of clinical significance. These results are similar to prior studies of executive functioning and psychosocial functioning in pediatric hydrocephalus, where significant differences were reported between hydrocephalus and typically developing groups on the BRIEF (a parent- and self-report measure of executive functioning) and BASC-3, but average T-scores were still generally less than 1.5 SD above the mean (Helder et al., 2011; Mahone et al., 2002; Zabel et al., 2011).

An important consideration when examining average scores in this population is the degree of variability within etiologies as well as the variability between etiologies. This can provide context for why an etiology group's mean scores may be in the average range despite

prior research indicating psychosocial difficulties in that group. An example of this is the myelomeningocele etiology group. Children with SBM have been reported to have higher levels of internalizing symptoms (Kelly et al., 2012) and difficulties with executive functioning and attention (Kelly et al., 2012; Swartwout et al., 2008) compared to typically developing controls. In the present study, the BASC-3 scores for the myelomeningocele group were all in the average range. Standard deviations may be as informative as average scores in a pediatric hydrocephalus population. For example, in the present study, average executive functioning T-score for the myelomeningocele group was 58, but the standard deviation was 9 (Table 2), meaning that 68% of communicating congenital hydrocephalus patients in the current study had executive functioning scores between 49 and 67. The mean score for depression was 57 with a standard deviation of 13. While this variability may make it difficult to directly compare etiology groups to each other, characterizing the general distributions of functioning in different etiology groups can help generate ideas about how best to evaluate and treat children with hydrocephalus. Most prior research on social-emotional functioning in pediatric hydrocephalus was limited to few etiologies, and often limited to just spina bifida (Helder et al., 2011; Jenkinson et al., 2011; Kulesz et al., 2015; Swartwout et al., 2008; Zabel et al., 2011).

Myelomeningocele and Psychosocial Functioning

Myelomeningocele was hypothesized to be the etiology group with greatest risk of psychosocial difficulties due to previous reports that it is a risk factor for worse overall HRQoL relative to other etiologies (Kulkarni et al., 2008b). It has also been found to be a risk factor for worse neuropsychological difficulties compared to aqueductal stenosis (Hampton et al., 2013; Swartwout et al., 2008), which was hypothesized to have an impact on psychosocial functioning. In the present study, the myelomeningocele group did not have significantly worse (higher)

BASC-3 scores than any other etiology. Myelomeningocele had significantly worse (lower) HOQ social-emotional scores than just one etiology group: the "other" group. It is possible that the medical comorbidities that often accompany SBM, such as mobility difficulties and neurogenic bowel and bladder do not impact psychosocial functioning as measured by the BASC-3 and HOQ to the extent expected.

In the present study, 43% of total participants responded "somewhat true", "quite a bit true", or "very true" to the HOQ item "My child has difficulty walking", and 27% responded "somewhat true", "quite a bit true", or "very true" to the HOQ item "My child needs a wheelchair." When just using the "quite a bit true" or "very true" responses to determine wheelchair use, 20% of the present study population appears to need a wheelchair. In the myelomeningocele group, these numbers are 89% who responded "somewhat true" or more to the question "My child has difficulty walking", 64% who responded "somewhat true" or more to the question "My child needs a wheelchair", and 46% who responded "quite a bit true" or "very true" that their child needs a wheelchair (Table 7). In a 2015 nationwide study of children with spina bifida, 37% of children with SBM were reported to require a wheelchair (were not community ambulators), and an additional 17% required a wheelchair sometimes but could ambulate at home or in a therapy setting (Dicianno et al., 2015). This 54% of reported SBM patients who required a wheelchair at times is slightly less than the 64% of children in the present study who appear to require a wheelchair at times.

Although there were not specific questions related to neurogenic bowel and bladder in the surveys used in the current study, the HOQ item "My child needs help using the washroom" can be examined as a proxy for bowel and bladder difficulties. In the overall study population, 40% responded "somewhat true", "quite a bit true", or "very true" to this item, compared to 82% of

the myelomeningocele group. When just looking at "quite a bit true" and "very true" responses, 35% of the overall study population endorsed that their child needs help with the washroom, compared to 71% of the myelomeningocele group (Table 7). Despite these mobility difficulties and potential bowel and bladder difficulties, the myelomeningocele group did not have worse parent-reported BASC-3 scores compared to other etiologies and they only had lower HOQ scores compared to the "other" etiology group.

Medical Risk Factors for Psychosocial Functioning (Hypothesis 2)

The second aim of this study was to examine medical risk factors for psychosocial functioning. Linear regression models with number of neurosurgical procedures, history of seizures, and years with hydrocephalus as predictor variables were significant for BASC-3 anxiety, attention, hyperactivity, and executive functioning scales and HOQ social-emotional scores but not for the BASC-3 depression scale. For the BASC-3 attention, hyperactivity, and executive functioning scales, years with hydrocephalus is the predictor variable driving this significant relationship. For every additional year with hydrocephalus, the attention problems scale is expected to increase by 1.06 points, the hyperactivity scale is expected to increase by 0.72 points, and the executive functioning scale is expected to increase by 1.24 points, holding the other variables constant (Table 3). For the BASC-3 anxiety scale, history of seizures was the only significant predictor, such that any history of seizures is associated with a 5 point reduction on the anxiety scale. This is an interesting result given that research on psychiatric comorbidities with epilepsy often reports higher rates of anxiety in children with epilepsy (Dagar & Falcone, 2020; Kwong et al., 2016; Schraegle & Titus, 2017) Many of these studies, however, compare children with epilepsy to typically developing controls rather than children with hydrocephalus. Future research may consider exploring potential mediators for this effect, such as medication or

cognitive functioning. For the HOQ social-emotional domain, history of seizures and years with hydrocephalus were significant predictors. Any history of seizures is associated with a -.09 point reduction (worse social-emotional health) on the HOQ and each additional year with hydrocephalus is associated with a -.01 point reduction. Number of neurosurgical procedures was not a significant predictor for any outcomes. Previous research examining number of procedures has reported variable findings regarding health outcomes. Kulkarni, Drake, and colleagues (2004) reported that number of shunt revisions in a mixed-etiology sample was significantly associated with cognitive and total HOQ score, but not with social-emotional score. Another study reported an association with physical HOQ score, but not cognitive or social-emotional score (Ros-Sanjuán et al., 2021). In later studies using the HOQ to measure health outcomes, number of hospital admissions was not associated with HOQ scores (Kulkarni et al., 2008b). In a study of children with hydrocephalus due to premature birth and intraventricular hemorrhage, number of neurosurgical procedures was not associated with HRQoL as measured by the PedsQL (Gigi et al., 2019). Although greater number of revisions may be associated with structural changes (Kulesz et al., 2019) and cognitive difficulties in children (Arrington et al., 2016), the current study indicates that number of CSF diversion surgeries is not a significant predictor of psychosocial health outcomes.

Although several of the regression models were significant, R^2 for each model was small, ranging from .08 for the anxiety and hyperactivity scales to .16 for the executive functioning scales. The medical variables examined here (number of procedures, history of seizures, and years with hydrocephalus) together only account for between 8 and 16% of the variance in BASC-3 and HOQ scores (Table 2). The coefficients for the significant variables tended to have wide confidence intervals. For example, although each additional year of hydrocephalus was

associated with an increase of 0.72 hyperactivity points, the 95% confidence interval was -0.01 to 1.45. Previous studies examining medical, social, and economic variables in relation to HOQ scores similarly found small R^2 for their regression models examining social-emotional and cognitive health outcomes. A higher proportion of variance in physical health outcomes was explained by their models. However, the present results replicate their finding that any history of seizures significantly impacts social-emotional HOQ scores (Kulkarni et al., 2008b). It is possible that other medical variables, such as comorbid medical conditions or family functioning (Kulkarni et al., 2008b) impact psychosocial functioning in the present population.

Given that some psychosocial difficulties may be more likely to present at different ages, regardless of hydrocephalus status, it is possible that age, rather than years with hydrocephalus, could be driving the significant relationship. In order to examine this possibility, exploratory analyses were conducted with the age at survey administration substituted for years with hydrocephalus in the regression models. This variable was not significant for any of the scales examined.

Socioeconomic Risk Factors for Psychosocial Functioning (Hypothesis 3)

The third aim of this study was to examine SES factors in relation to psychosocial functioning. Parent education (some high school, completed high school, completed 2-year degree, completed 4-year degree, completed master's degree, completed doctorate degree), income ($0-$20,000, $20,000-$40,000, $40,000-$60,000, $60,000-$80,000, $80,000-$100,000, >$100,000), and employment (at least one parent full time employed, no parent full time employed) were included as predictors in linear regression analyses examining BASC-3 anxiety, depression, attention, hyperactivity, and executive functioning and HOQ social-emotional scores as outcomes. Regression models with parent education, income, and employment were not

significant for any psychosocial outcome examined. This highlights that although SES has been linked to health outcomes and reported HRQoL in children (Kulkarni, 2010), SES may not always have a straightforward link to psychosocial outcomes. Indeed, several previous studies using the HOQ did not report an association between SES variables and social-emotional functioning. One study found no association between insurance as a proxy for SES and overall HOQ scores (Reynolds et al., 2020), while another found an association of household income with overall HOQ scores but not with social-emotional scores (Karmur & Kulkarni, 2018), similar to our results.

It was somewhat surprising, however, that BASC-3 measures of psychosocial functioning were not associated with SES variables. A systematic review of studies from 1990 to 2011 found that SES, as measured by at least one of: household income, poverty, parental education, parental occupation status, or family affluence, was associated with higher rates of mental health problems (Reiss, 2013). That review found that low SES that persisted over time was more strongly related to higher rates of mental health problems. It is important to note that the current study captures one point in time for SES variables, such that it is impossible to know from the current data if the reported SES is stable or temporary, which may impact the degree to which our data could capture longstanding SES status.

Additionally, in that review, household income and parental education were some of the most consistent predictors of mental health problems. Many of the studies reviewed in that paper and in other studies of SES and health outcomes have a broader range of SES statuses, including a larger number of patients living in poverty or with very low education (Poulain et al., 2020; Reiss, 2013). In the present study population, at least one parent had completed high school in each family who participated, and 49% of the adults reported on had a bachelor's degree,

compared with 33% of the general US population (U.S. Census Bureau, 2020). Additionally, in the present study, 50% of respondents had a family income of $80,000 or above, and 68% had a family income of $60,000 or above. The median U.S. household income in 2020 was $67,521 (U.S. Census Bureau, 2020). An additional factor to consider when measuring SES is that families with low SES tend to have worse health status and less access to medical care (Newacheck et al., 1996). It is possible that the families more likely to agree to participation in this study were also those with more regular medical care and higher SES. The relatively high level of income and education in this population may have made it difficult to detect the full impact of SES on psychosocial outcomes.

An additional component of SES that was not considered in the present study is race and ethnicity. For race, 94% of respondents were classified as “white”, 1.6% as “Asian”, 1.6% as “Black or African-American”, and 1.6% as “American Indian or Alaska Native.” For ethnicity, 87% or respondents were classified as “non-Hispanic or Latino”, with 13% classified as “Hispanic or Latino”, according to electronic medical records. The constrained range of race and ethnicity in the study population made it a difficult variable to include as a component of SES, despite an important body of research identifying race and racism as important mental health outcome predictors (Bailey et al., 2017; Berger & Sarnyai, 2015; Paradies et al., 2015).

Since race is not a variable collected in the Hydrocephalus Clinical Research Network Registry, it is unknown if these numbers are representative of the racial and ethnic distribution of all hydrocephalus patients at Primary Children’s Hospital. The racial and ethnic distribution in the present study does vary from the general population, which is estimated to be 76% white, 13% Black or African American, 1.3% American Indian or Alaska Native, 6% Asian, and 0.2% Native Hawaiian or other Pacific Islander (U.S. Census, 2020). The non-significant results for

this hypothesis are a reminder that, although SES is a well-established factor in health outcomes, the specific SES variables examined and the study population can have an impact on the relationships observed.

Limitations and Future Research

One limitation of the present study was the lack of information about cognitive functioning or IQ as a variable. While the HOQ includes a cognitive functioning scale, there are only 12 questions and they are not very specific regarding cognitive domains (Kulkarni, Rabin, et al., 2004). The goal of the present study was to include a wide range of children with hydrocephalus to establish a broad understanding of psychosocial functioning in this population. Low cognitive functioning, however, can certainly impact psychosocial functioning (Lindquist et al., 2006) and is likely an important consideration in understanding the psychosocial variables examined here. In particular, executive functioning and attention problems may correlate with cognitive difficulties in this population. However, prior research that has controlled for IQ has still found increased levels of psychosocial difficulties in this population compared to health controls (Helder et al., 2011). Future research on psychosocial functioning in pediatric hydrocephalus may expand our understanding of how cognitive difficulties can have an impact.

An additional limitation of the study was the lack of a medical control group with which to compare the pediatric hydrocephalus group. Scores were instead compared to the BASC-3 normative data, but this data comes from a different population than the population the study sample came from. The present study also only included English-speaking participants. While this was a necessary choice due to limited reliability and validity data regarding the Spanish-language HOQ and lack of availability of the HOQ in other languages, this does limit ability to generalize results to a broader, non-English speaking population. It also could have created

constraints on the distribution of SES variables. As discussed, the present study sample had higher than average income and education levels. It additionally did not include race as an SES variable. Future research may elect to expand SES variables in order to gain a better understanding of how SES impacts this population.

The primary goal of the present study was to include all etiologies of hydrocephalus to expand on prior research that was often limited to one or two etiologies. However, we were unable to separate etiology groups into degree of severity within the etiology, which could be a component of understanding psychosocial functioning within etiologies. For example, it has been observed that the severity or comorbidities within an etiology significantly contributes to outcomes, as in spinal lesion level in myelomeningocele (Fletcher et al., 2005) or the presence of cerebral palsy in hydrocephalus due to IVH secondary to prematurity (Gigi et al., 2019). Future research may elect to include all etiologies as well as outcome factors within etiologies, knowing this will provide information for a wider range of patients.

The present results vary somewhat from a recent study that reported no differences between pediatric hydrocephalus patients and the normative population in self-reported anxiety and depression (Zimmerman, May, Barnes, Arynchyna, Alford, Wessinger, et al., 2020). This is perhaps not surprising, as children with disabilities have been observed to report their own quality of life as similar to that of typically developing peers, despite their extent of impairment (Feingold et al., 2002; Mezgebe et al., 2015; Zwicker & Harris, 2008). This could extend to self-report of psychiatric symptoms. While the present study population was not an appropriate population for self-report due to young age and degree of impairment of some participants, future research on psychosocial functioning in pediatric hydrocephalus may consider including both self- and proxy-report in order to further understand how this population perceives their

own functioning compared to how they are seen by others. This can have important treatment implications, as psychotherapy for children and adolescents often includes the family.

Conclusions

This study found that hydrocephalus etiology impacts psychosocial functioning, as measured by the BASC-3 executive functioning and attention scales and the HOQ social-emotional scale. Etiology groups did not differ on BASC-3 anxiety, depression, or hyperactivity scales, indicating that other variables may be more relevant for psychosocial outcomes. Medical factors, including history of seizures and years with hydrocephalus, were found to be significant predictors for BASC-3 anxiety, attention, hyperactivity, executive functioning scales and HOQ social-emotional scale, but not for BASC-3 depression. Effect sizes for etiology and amount of variance explained by medical predictors were small. This is likely partially due to the variability in functioning in a pediatric hydrocephalus population that includes all etiologies. Socioeconomic variables were not found to be significant predictors of psychosocial outcomes. This research provides information regarding psychosocial outcomes for children with hydrocephalus of etiologies. It also provides a framework for future research attempting to identify important factors and interventions for psychosocial health in this population.

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Table 1

Patient Characteristics

	Participants (n = 125)	Non-Participants (n = 272)
Age in yrs, mean (SD, range)	9.8 (3.1, 5.2-17.9)	10.5 (3.1, 5.1-17.9)
Sex, n (%)		
Female	48 (38)	114 (42)
Male	77 (62)	158 (58)
Etiology, n (%)		
Post-IVH second to pre.	22 (18)	59 (22)
Myelomeningocele	28 (22)	51 (19)
Comm. Congenital	12 (10)	17 (6)
Aqueductal Stenosis	13 (10)	28 (10)
Other	50 (40)	117 (43)
Age at first permanent CSF diversion surgery in days, mean (SD, range)	914 (1443, 1-5557)	731 (1255, 1-5783)
Total number of CSF diversion surgeries, mean (SD, range)	2.4 (2.1, 1-16)	2.3 (2.2, 1-22)

Table 2

Kruskal-Wallis of BASC-3 and HOQ Scores by Etiology

Scale	Etiology					χ^2	η^2	*p*
	Post-IVH Secondary to Prematurity *(n = 22)*	Myelo. *(n = 28)*	Comm. Congenital *(n = 12)*	Aqueductal Stenosis *(n = 13)*	Other *(n = 50)*			
BASC-3 Anxiety* (n = 123)	50 (12)	55 (12)	49 (11)	53 (15)	53 (11)	2.61	.02	.62
BASC-3 Depression (n = 123)	56 (10)	57 (13)	55 (10)	54 (9)	56 (12)	0.80	.007	.94
BASC-3 Attention (n = 123)	63 (9)	55 (8)	60 (11)	53 (11)	55 (12)	10.28	.08	**.04**
BASC-3 Hyperactivity (n = 123)	60 (16)	55 (11)	57 (16)	50 (6)	55 (14)	3.25	.03	.52
BASC-3 Executive Functioning (n = 122)	66 (9)	58 (9)	61 (15)	56 (11)	57 (13)	10.55	.09	**.03**
HOQ Social-Emotional (n = 125)	.62 (.18)	.66 (.19)	.70 (.21)	.74 (.14)	.75 (.18)	10.57	.08	**.03**

**BASC-3 scores have a mean of 50 and SD of 10*
***HOQ scores range from 0.0 (worse health) to 1.0 (better health) with a difference of 0.10 being clinically meaningful*
Note: Bolded text indicates statistical significance

Table 3

Multiple Regression Analyses of Medical Variables

Scale	*t*	*p*	B (95% CI)	*F*	*df*	*p*	R^2
BASC-3 Anxiety* (n = 123)							
Overall Model				3.30	3, 119	**.02**	.08
Procedures	-1.75	.08	-0.90 (-1.92, 0.12)				
Seizures	-2.10	**.04**	-5.43 (-10.54, -0.31)				
Years with Hydro	1.01	.31	0.34 (-.33, 1.01)				
BASC-3 Depression (n = 123)							
Overall Model				1.57	3, 119	.20	.04
Procedures	-0.06	.96	-0.03 (-1.01, 0.95)				
Seizures	-0.75	.54	-1.87 (-6.79, 3.06				
Years with Hydro	2.06	**.04**	0.67 (0.03, 1.31)				
BASC-3 Attention (n = 123)							
Overall Model				5.36	3, 119	**.002**	.12
Procedures	0.05	.96	0.02 (-0.89, 0.94)				
Seizures	1.51	0.13	3.51 (-1.08, 8.09)				
Years with Hydro	3.48	**.001**	1.06 (0.46, 1.66)				
BASC-3 Hyperactivity (n = 123)							
Overall Model				3.38	3, 119	**.02**	.08
Procedures	1.12	.26	0.63 (-0.48, 1.73)				
Seizures	1.54	.13	4.32 (-1.24, 9.88)				
Years with Hydro	1.96	**.05**	0.72 (-0.01, 1.45)				
BASC-3 Executive Functioning (n = 122)							
Overall Model				7.61	3, 118	**<.001**	0.16
Procedures	1.01	.31	0.49 (-0.47, 1.45)				
Seizures	1.58	.12	3.86 (-0.97, 8.69)				
Years with Hydro	3.86	**<.001**	1.24 (0.60, 1.87)				
HOQ Social-Emotional (n = 125)							
Overall Model				5.37	3, 121	**.002**	.12
Procedures	-0.94	.35	-.007 (-.023, .008)				
Seizures	-2.26	**.03**	-.09 (-.16, -.01)				
Years with Hydro	-2.51	**.01**	-.01 (-.02, -.003)				

**BASC-3 scores have a mean of 50 and SD of 10*

***HOQ scores range from 0.0 (worse health) to 1.0 (better health) with a difference of 0.10 being clinically meaningful*

Note: Bolded text indicates statistical significance

Table 4

Multiple Regression Analyses of SES Variables

Scale	*F*	*df*	*p*	R^2
BASC-3 Anxiety* (n = 123)	0.73	10, 111	.70	.06
BASC-3 Depression (n = 123)	0.50	10, 111	.89	.04
BASC-3 Attention (n = 123)	1.08	10, 111	.39	.09
BASC-3 Hyperactivity (n = 123)	0.49	10, 111	.89	.04
BASC-3 Executive Functioning (n = 122)	0.90	10, 110	.53	.08
HOQ Social-Emotional (n = 124)	0.61	10, 113	.80	.05

**BASC-3 scores have a mean of 50 and SD of 10*
***HOQ scores range from 0.0 (worse health) to 1.0 (better health) with a difference of 0.10 being clinically meaningful*
Note: Bolded text indicates statistical significance

Table 5

At Risk (T ≥ 60) or Clinically Significant (T ≥ 70) BASC-3 Scores

BASC-3 Scale	% At Risk	% Clinically Significant
Clinical Scale		
Hyperactivity	14	19
Aggression	19	12
Conduct	13	10
Anxiety	13	7
Depression	21	9
Somatization	18	12
Atypicality	22	24
Withdrawal	20	13
Attention	25	16
Adaptive Scale		
Adaptability	25	5
Social Skills	19	11
Leadership	43	14
Activities of Daily Living	29	24
Functional Communication	28	22
Content Scale		
Executive Function	33	19

Note: In a typically developing population, 13.6% of scores are expected to be "at risk" and 2.3% of scores are expected to be "clinically significant"

Table 6

BASC-3 Scores: Children with Hydrocephalus Compared to Normative Sample

BASC-3 Scales	Patient Scores	*t*	Cohen's *d*	*p*
Anxiety	53 (12)	2.48	.22	.014
Depression	56 (11)	5.71	.51	<.0001*
Hyperactivity	55 (13)	4.63	.42	<.0001*
Attention	57 (11)	6.70	.61	<.0001*
Executive Functioning	59 (12)	8.41	.76	<.0001*

**p < .01, Bonferroni correction for 5 comparisons*
Note: one sample T-tests compared to BASC-3 normative sample with T-score of 50

Table 7

Responses for Select HOQ Items

		Etiology				
		Post-IVH Secondary to Prematurity *(n = 22)*	Myelo. *(n = 28)*	Comm. Congenital *(n = 12)*	Aqueductal Stenosis *(n = 13)*	Other *(n = 50)*
Quite a bit true and very true	My child needs help using the washroom	55%	71%	8%	31%	14%
	My child has difficulty walking	36%	64%	8%	23%	16%
	My child needs a wheelchair	23%	46%	8%	15%	8%
Somewhat true, quite a bit true, and very true	My child needs help using the washroom	55%	82%	17%	38%	16%
	My child has difficulty walking	55%	82%	25%	38%	22%
	My child needs a wheelchair	27%	64%	17%	15%	12%

Table 8

Mean HOQ Scores Across Publications

HOQ scale	Present study	(Kulkarni, Drake, et al., 2004)	(Kulkarni, Cochrane, McNeely, et al., 2008)	(Zimmerman, May, Barnes, Arynchyna, Alford, Chagoya, et al., 2020)
Total	.64	.68	.65	.68
Physical	.63	.71	.66	.69
Cognitive	.54	.57	.55	.54
Social-emotional	.70	.72	.71	.73