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Demographics and Characteristics of Hydrocephalus in Adults: The First 500 Subjects of the Adult Hydrocephalus Clinical Research Network Registry

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AHCRN
Adult Hydrocephalus Clinical Research Network

INTRODUCTION

The characteristics of the adult population with hydrocephalus are not well described. The Adult Hydrocephalus Clinical Research Network (AHCRN), founded in 2014, initially comprised University of Calgary, University of British Columbia, Cleveland Clinic, Weill Cornell Medical College, Sinai Hospital of Baltimore, University of Washington, and University of Utah (Data Coordinating Center).

LEARNING OBJECTIVES

By the conclusion of this session, participants should be able to describe the range of adults with hydrocephalus, including categories and etiologies

METHODS

Adults >=18 years were non-consecutively enrolled in a registry. Data includes symptoms, examination, neuropsychology screening, comorbidities, imaging, treatment, complications, and outcomes.

Four categories were defined:
Transition (treated before age 18),
Congenital (congenital pattern, not treated before age 18),
Acquired (secondary to known risk factors, treated or untreated),
Possible Idiopathic Normal Pressure Hydrocephalus (>=age 65 years; referred for evaluation of INPH; not previously treated).

We report the first 519 subjects (2015-2017).

RESULTS

Demographics
-Sex: Female 42.0%
-Age at Enrollment: 59.8 ± 19.2 years
(Range 18.1 - 90.7)
-Race: White 90.6%, Asian 5.0%, Black 1.7%, Missing/Other 2.7%

Category
-Transition: n=86 (16.6%)
-Congenital: n=132 (25.4%)
-Acquired: n=88 (17.0%)
-Possible INPH: n=213 (41.0%).

Category Variation Among 6 Centers:
-Transition: 0-43%
-Congenital: 0-42.2%
-Acquired: 8.7-31.9%
-Possible INPH: 19.0-85.7%.

Treatment at Time of Enrollment
-All Subjects: 36.8% (Shunt 26.8%, ETV 10.0%)
-Transition: 100% (Shunt 95.3%, ETV 14.0%)
-Congenital: 43.2% (Shunt 22.0%, ETV 21.2%)
-Acquired: 39.2% (Shunt 27.5%, ETV 11.8%)
-Possible INPH: 0% (By definition)

Top Etiologies
-Transition: myelomeningocele 33.7%, aqueductal stenosis 15.1%, IVH of prematurity 9.3%
-Congenital: communicating 41.7%, aqueductal stenosis 50.0%, aqueductal pattern 15.2%
-Acquired: Brain tumor 28.4%, intraventricular adhesion/web/colloid cyst 13.7%, non-traumatic SAH 11.8%, TBI 9.8%

Lawton ADL Score (lower=better)
-Transition: 3.1 ± 4.0
-Congenital: 1.5 ± 2.7
-Acquired: 4.1 ± 5.9
-INPH: 5.5 ± 5.2.

CONCLUSIONS

The population in the AHCRN registry is diverse. Because the centers in the AHCRN are all academic specialty centers, and because the enrollment in the registry was non-consecutive, the results and trends identified may or may not reflect the broader population of adult hydrocephalus in the US and Canada.

A combined 42% of enrolled subjects have childhood onset hydrocephalus (whether treated or untreated by age 18), i.e, Transitional and Congenital categories. Only 41% of enrollees were seen for possible INPH.

The proportion of hydrocephalus categories varied widely among centers. Some centers are primarily focused on INPH, while others serve all 4 adult hydrocephalus cetegories. The center differences, and the registry as a whole, show that the population of adults with hydrocephalus is not restricted to iNPH or acquired hydrocephalus. Patients in the Transition category face the challenge of leaving the pediatric health care system to enter the adult health care system to receive longitudinal care.

The degree of impairment (Lawton ADL) is least in the Congenital group and worst in possible INPH, but all groups have significant degrees of impairment and disability. Because of the lifetime risk of treatment failure for hydrocephalus, adults with hydrocephalus need periodic evaluations to determine whether changes in their symptoms, neurologic examination, or brain imaging reflect hydrocephalus or other comorbidities.

More research on the lifespan health care needs and outcomes for all categories of adult hydrocephalus is needed.

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