

Neurocognitive Impairment is Common in the Adult with Congenital Heart Disease: Identification using a novel clinical questionnaire

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Background: Studies have found that children with congenital heart disease (CHD) and adults with acquired heart disease are at an increased risk of neurocognitive impairment. The prevalence of neurocognitive impairment and its risk factors in the adult congenital heart disease (ACHD) population is not well described.

Methods: The Wisconsin Adult Congenital Heart Disease (WAtCH) Program recently began screening patients to identify those at high risk of having neurocognitive impairments. Patients without congenital heart disease, a known genetic syndrome, or a previously diagnosed neurocognitive disorder were excluded from the screening process. Screening consists of using a validated neuro-oncology screening instrument that has been modified for the ACHD population. It consists of 10 questions answered on a Likert scale that measure a subject's self perceived competence in 8 neurocognitive domains (attention, memory, language, executive functioning, independence in activities of daily living, math, processing speed and reading). Those patients who answer this survey in a predetermined high risk fashion for a neurocognitive disorder are then referred for a formal neurocognitive evaluation. A database consisting of the patients that are screened, those that meet criteria to be referred and the answers to the questions has been developed. Demographic information, type of CHD, number of surgeries and medications at the time of the survey are obtained by chart review.

Results: In the pilot phase of the neurocognitive screening program 182 patients (43% males) were eligible and completed the neurocognitive screening process. The average age of patients who underwent screening was 31 years (range: 18-69 years) For the cohort, 33 (18%) patients had not had a prior cardiac surgery, 54 (30%) one surgery, 54 (30%) two, and 41 (22%) at least three. Out of the 182 patients screened, 60 (33%) met the high risk criteria to be referred for a formal neurocognitive evaluation. Patients who met the high risk criteria were older (33 years vs 30 years, $p=0.028$), had undergone a greater number of prior cardiac surgeries (2.1 vs 1.5, $p=0.008$), had more complex CHD ($p=0.026$) and longer duration of cyanosis (9.6 years vs. 2.5 years, $p=0.007$). Of those patients that met the criteria of being high risk, the neurocognitive domains in which they had the worst perceived functioning were attention and math. The domain in which they had the best perceived functioning was independence in activities of daily living.

Conclusion: Our pilot data, confirms that neurocognitive impairment is common in ACHD patients and in many cases under recognized. Simple screening questionnaires may help identify those patients at high risk and allow for timely and appropriate referral for formal neurocognitive evaluation, diagnosis and therapy. Prospective validation of the screening survey with results of formal neurocognitive evaluation in adult CHD population is currently ongoing.