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ANESTHESIA MANAGEMENT FOR PEDIATRIC PATIENTS WITH HIGH RISK WILLIAMS SYNDROME - A QUALITY IMPROVEMENT STUDY

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Introduction:

Over 80% of patients with Williams syndrome (WS) have structural cardiovascular abnormalities[1], predisposing them to a risk of adverse cardiac events (ACE) during general anesthesia [2,3]. Intravenous induction of anesthesia is usually preferred for high risk patients however, mask induction with volatile anesthesia is sometimes necessary especially in younger patients and those with difficult intravenous access when they present on the day of intervention. In 2017 there was a change in anesthesia management at our institution which is a national referral center for WS. Based on a 3-staged risk category (RC) for WS patients[4], patients are admitted for intravenous pre-hydration the evening before (high RC), 2 hours prior to anesthesia (moderate RC) or they received standard care (low RC). The aim of this quality improvement study was to evaluate if a change in anesthesia management based on risk stratification and pre-operative intravenous hydration and intravenous induction decreased the incidence of ACE during anesthesia induction in WS patients.

Methods:

After IRB approval, we identified all pediatric patients with WS who underwent anesthesia for catheterization lab procedures, cardiovascular or non-cardiovascular surgery or diagnostic imaging between 11/2008 and 08/2019. We reviewed electronic anesthesia records and compared the intervention group (IG, 03/17 – 08/19) to a historical control group (HG, 11/08 – 02/17). Primary outcome were ACE (defined as cardiac arrest, CPR, arrhythmias or ST-segment changes) within 60 minutes after anesthesia induction. Secondary outcome were more than two inotrope bolus given and the percentage change in the systolic blood pressure during anesthesia induction. Standardized mean difference (SMD) was calculated, with a SMD >0.2 suggesting clinically significant difference between the groups.

Results:

We identified 142 patients with WS, of these 48 underwent 118 anesthesias. In the HG 27 patients had 67 anesthesias, and 28 in the IG had 51 anesthesias. The incidence of ACE in the HG was 6% vs 2% in the IG (SMD=0.207). The frequency of more than two inotrope bolus given was 13.4% in the HG vs 11.8% in the IG (SMD=0.069) and the median drop of the systolic blood pressure was 17.5% (IQR: 5-30%) in the HG vs 9% (IQR: 5-18%) in the IG (SMD=0.419).

Conclusion:

This is the first quality improvement study investigating an adapted anesthesia management strategy in WS patients with regard to ACE during anesthesia induction. The results suggest a clinically significant reduction of ACE and a more stable systolic blood pressure during anesthesia induction in patients who received pre-operative intravenous hydration and intravenous induction. The presence of intravenous access allowed for titration of induction drugs, an immediate response to arterial hypotension and the pre-induction initiation of inotropic support. The presented risk

stratification for intravenous access and an adapted anesthesia management should be considered in WS patients.

References:

1 Collins RT. Cardiovascular disease in Williams syndrome. Circulation. 2013;127:2125-34.

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3 Burch TM et al. Congenital supravalvular aortic stenosis and sudden death associated with anesthesia: what's the mystery? Anesthesia and analgesia. 2008;107:1848-54.

4 Collins RT et al. Peri-procedural risk stratification and management of patients with Williams syndrome. Congenital heart disease. 2017;12:133-42.