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SAFETY AND EFFICACY OF LOW DOSE DIAZOXIDE FOR TREATMENT OF HYPERINSULINEMIC HYPOGLYCEMIA IN SMALL-FOR-GESTATIONAL AGE INFANTS

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Background The use of Diazoxide (DZX) among small for gestational age (SGA) infants with hyperinsulinemic hypoglycemia (HH) has risen in the last decade. Since the aetiology of HH in SGA is primarily multifactorial and non-genetic, DZX is an effective first-line medication. However, in therapeutic doses (5–15mg/kg/day), DZX has potentially serious adverse effects such as pericardial effusion, pulmonary hypertension and neutropenia. Yet, the literature on the safety and efficacy of low dose DZX (<5mg/kg/day) in SGA infants is lacking.

Objectives To assess the safety and efficacy of low dose DZX for the treatment of HH in SGA infants.

Methods This is a retrospective study of SGA infants with HH treated with DZX between 1st September 2014 and 31st September 2020. Neonates with sepsis, severe perinatal asphyxia or contraindications to feeding were excluded. DZX was initiated at a dose of 3mg/kg/day in 2 divided doses (with Hydrochlorothiazide) among HH infants with suboptimal response to rising glucose infusion rates (GIR). DZX dose was increased to 5mg/kg/day, and then in increments of 2.5mg/kg/day if required. Pre-requisites for starting DZX include an echocardiogram for pulmonary hypertension and pre-existing pericardial effusion, normal liver and renal function tests. Feeds are escalated and GIR weaned when pre-feed glucose levels demonstrate DXZ response. Safety fast study was done prior to discharge home on DZX and all infants continued with home glucose monitoring. DZX was discontinued when doses self-weaned to <1.5mg/kg/day with weight gain. A resolution fast study (RFS) was conducted after 72 hours without DZX.

Results Of 57 SGA infants with HH, 27 (47%) required DZX treatment, while the rest achieved spontaneous resolution. Among DZX treated infants, 15 (55%) were male, 12 (45%) were preterm, mean gestational age was 36.4 ± 2 weeks, and birth weight was 1942 ± 356 grams. Five infants presented with jitteriness (18.5%) and one with seizures (3.7%) and they presented at a mean of 1.1 ± 0.4 days of life. The mean paired values of glucose and insulin were 2.37 ± 0.47 mmol/l and 16.4 ± 27.4 mU/l, respectively. Diazoxide was initiated at a mean of 13 ± 8 days of birth and the mean dose required was 4.6 ± 2.2 mg/kg/day, with 89% receiving ≤ 5 mg/kg/day. The mean duration of DZX treatment was 66 ± 40 days. Subgroup analysis showed that initiation of DZX <10 days of life led to earlier resolution of HH (11.1 ± 2.8 vs 20.6 ± 6.6 days; $p < 0.001$), shorter duration of central line use (11.3 ± 3.5 vs 19.6 ± 4.8 days; $p < 0.001$) and shorter hospital stay (18.3 ± 6.7 days; $p = 0.022$), compared to those who started DZX after 10 days. Two infants (7.4%) who required 10mg/kg/day had hypertrichosis and one infant (3.7%) on 4.8mg/kg/day had fluid retention and oedema. Upon discontinuation of DZX, 93% passed a formal hospital-based RFS.

Conclusions Low dose DZX is safe and effective for the treatment of HH in SGA infants. Even at low doses, treatment was effective and short term side effects were uncommon. However, being a K_{ATP} channel agonist, there is potential for DZX to cause neuronal hyperpolarization leading to brain injury. Future larger cohort studies will help to determine the long-term outcome of infants treated with DZX, especially on higher doses.

Paediatricians with Expertise in Cardiology Special Interest Group

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OUTCOME OF CHILDREN WITH FAMILY HISTORY OF CONGENITAL OR INHERITED CARDIAC CONDITIONS REFERRED TO PAEDIATRIC CARDIOLOGY CLINICS

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Background About 1% of children are born with congenital heart disease (CHD). Inherited cardiovascular conditions (ICC) including cardiomyopathies and inherited arrhythmias are rare but can be associated with increased mortality and morbidity. The risk of developing an ICC or CHD rises significantly in children with a positive family history (FHx).

Objectives

1. To determine the outcome of children seen with a family history of CHD or ICC at Cambridge University Hospitals Foundation Trust (CUHFT)
2. To review the referral pathway for children seen in the local paediatric cardiology services and develop a shared care approach to on-going care with clinical geneticists and the specialist ICC service.

Methods Retrospective review of the hospital records of all children under 17 with a FHx of CHD or ICC who were seen by Paediatricians with Expertise in Cardiology (PECs) since 2015 with a comprehensive review of patients seen in 2019 in a busy level 3 Local Children's Cardiology Centre. Patients were identified from the EPIC medical records by specific diagnosis reports.

Results There has been a steady increase in the number of patients attending clinics from 173 in 2015 to 224 in 2019. In 2019, 127(57%) were new patients while 97(43%) were follow-up. New referrals were seen at a median age of 7 months, range 2 weeks to 16.6 years. 101 (45%) were male and 123 (55%) female.

126 (56%) were referred for investigation of FHx of CHD, 52 (23%) with FHx of cardiomyopathy, 20 (9%) for FHx of cardiac arrhythmias, 11 (5%) with a FHx of sudden death and 15 (7%) were seen for FHx of other conditions that could not be classified as CHD or ICC. 17/224 (8%) were referred from the maternity unit or the GP clinic with insufficient data about family history categorised as a hole in the heart, unspecified heart conditions and leaky valves that had not required any intervention.

98 (44%) infants had their first clinic assessment before 2 months of age; of these 36 (37%) had a PFO that required follow-up. 152/224(68%) children had normal echocardiography, whilst 72 (32%) had echocardiographic findings, half (36) had PFO, 8 had ASD, 8 had Bicuspid Aortic Valve, 5