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DILATED CORONARY SINUS IN NEONATES

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Background The coronary sinus is a venous system situated at the back of the atrioventricular junction and draining into the right atrium, which can be assessed from various echocardiography angles. In certain cases, the coronary sinus can be dilated by volume or more rarely by pressure overload.

Objectives To evaluate the efficacy of detecting coronary sinus in neonates and to determine the standard values of coronary sinus measurements. To highlight the importance of functional echocardiography in the assessment of the causes of dilation.

Methods Screening with echocardiography was done in 30 neonatal patients who were admitted to our unit (median 34-week gestation). Coronary sinus was evaluated in the four-chamber view (83% of cases), subcostal view (8%) or parasternal short-axis view (12%). The second group of two patients with a dilated coronary sinus was compared with the normal group.

Results Adequate coronary sinus imaging was done in 97.4 per cent of normal neonates. The diameter of the coronary sinus varied from 1 mm to 3.4 mm. The two neonates with a dilated coronary sinus had a persistent left superior vena cava, which flowed into it. In the abnormal group, the diameter of the coronary sinus was nearly three times greater. CS should be assessed and indexed to the body surface area as part of standard echocardiography screening. The dilated coronary sinus (+2 Z score) has several causes, including persistent left superior vena cava in our case, a total anomaly in the pulmonary venous drainage, pathological hepatic venous drainage or sinus ASD. A thorough evaluation of the Right Ventricle (RV) function is required, as dilated CS is one of the characteristics of RV dysfunction.

Conclusions Dilated CS can be contributed by many factors and detailed imaging is recommended, in a few patients, a contrast echo can help in visualising in details CS and rule out deroofting.

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CROUCH GAIT IN DRAVET SYNDROME: CLINICAL, PRACTICAL AND PERSONAL OBSERVATIONS

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Background Dravet syndrome (DS) is an aggressive developmental and genetic epileptic encephalopathy. Along with pharmacoresistant epileptic seizures, this rare disease is characterised by cognitive impairment, speech and language disability and, notably, mobility problems. Observational reports have shed some light on the progression of the disease, but the aetiology of DS is still poorly understood. The syndrome is further characterised by a progressive non-specific decline in gait. There is limited literature about the natural history of progressive gait decline in the context of DS.

Objectives In order to explore DS and its significant impact on patients' and their families, this work will discuss personal experiences caring for a patient with this condition, followed by a systematic literature review focusing on the aetiology and natural history of gait abnormalities in DS, with reflections regarding the aforementioned case.

Abstract 93 Table 1 Table showing the key studies utilised for this literature review.

n	Source of Journal Article	Lead Author, Year of Publication	Title	Number of participants
1	PubMed	Black, 2016	Crouch Gait and Dravet Syndrome	12
2	PubMed	Di Marco, 2019	Gait abnormalities in people with Dravet Syndrome: A cross sectional multi-centre study	71
3	PubMed	Gitiaux, 2016	Motor neuropathy contributes to coughing in patients with Dravet Syndrome	12
4	PubMed	Rilstone, 2012	Dravet syndrome: seizure control and gait in adults with different SCN1A mutations	10
5	PubMed	Rodda, 2012	Progressive gait deterioration in adolescents with Dravet Syndrome	26
<i>Total: 131</i>				

Methods The search via PubMed produced a total of nine articles. Four articles were excluded from the in-depth literature review as they focused on treatment options for patients with DS rather than the impact on crouch gait in children with DS.

Results The total number of children studied in this systematic review is 131. The final five articles are listed in table 1.

The literature reveals that children with DS show progressive gait deterioration that is noticeable in early childhood but has particular impact in the second decade of life. The literature suggests that children under 6 have a normal or variable gait pattern with possible features of joint hypermobility. Beyond this age, the progressive decline in gait is more heterogeneous. The literature indicates that there may be an evolution element to the gait deterioration that requires further investigation.

Over the course of six years caring for Child X, I have witnessed a serious visible decline in their mobility. The change has been gradual but the impact this has had on their independence is massive. Child X follows a classic trajectory of progressive gait deterioration, as observed by the majority of authors in more than half of the population cohort, in the literature review.

Conclusions Caring for a young child with DS, has provided me with a unique insight and first-hand account of the deterioration of their mobility, as well as the impact this has on their quality of life. More consideration is now being directed towards how families cope in their daily lives with the multiple aspects of DS.

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EVALUATION OF USE OF TECHNOLOGY AND ITS IMPACT ON GLYCEMIC CONTROL IN CHILDREN & YOUNG PEOPLE WITH TYPE1 DIABETES MELLITUS

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Background Technology in Diabetes has been rapidly evolving in order to help optimize blood glucose (BG) control in diabetic patients.

Objectives Analysis of glycemic control (GC) in children and young people with Type 1 Diabetes Mellitus (CYP T1D) using

Insulin Pump(IP) compared to those on Multiple Daily Injection(MDI) insulin regimen and assess if modality of BG monitoring has an impact on GC. To further assess the correlation of GC with age of the patient. Secondary outcomes included severe hypoglycemia and DKA episodes.

Methods Retrospective data was collected from FilePromaker database from June 2019 to October 2020 for all CYP T1D under the care of the Paediatric Diabetic Unit. This included data on age, management regime, modality of BG monitoring, HbA1c (laboratory and validated point of care blood tests at each diabetes related appointment), Diabetic Ketoacidosis (DKA) readmissions and severe hypoglycemia episodes. Mean HbA1c(mHb) was calculated for each patient during the study period in mmol/mol. GC was compared in two groups IP and MDI. Further analysis to evaluate impact of modality used for BG monitoring was undertaken. This included finger prick self blood glucose monitoring (SBGM) with a smart BG meter, flash glucose monitoring (FGM) or continuous glucose monitor (CGM). We also further looked at GC for children under 12 years(y) and over 12y in each group.

Abstract 94 Table 1 Insulin Delivery modality vs BG Monitoring modality vs Mean HbA1C

Mean HbA1C (mmol/mol)							
MDI(143)	CGM		FGM		SBGM		
	NO.		NO.		NO.		
<12	13	64.5	21	64.9	11	61.5	
>12	16		73.2	34	78.4	48	72.1
Overall	29		55		59		
IP (98)							
<12	25	56.1	8	63.9	0		
>12	20		60.3	22	66	23	64.4
Overall	45		30		23		

Results

- Total CYP T1D included in study were 241.
- MDI users were 143(60%) and IP users were 98(40%).
- Mean HbA1C in mmol/mol in IP group was 61.9(SD 10.23) & MDI group was 71.1(SD 17.34) which was statistically significant (t value of -4.74 and significance $p < 0.00$).
- GC overall in <12y vs >12y with mHb 60.98 vs 69.09($t = -58.91, p < 0.00$).
- In MDI group: mHb was highest in patients on FGM >12y=78.4; & lowest in those on SBGM <12y=61.5.
- However in Patients on IP: those on CGM and <12y had the lowest mHb of 56.1 and highest in those using FGM >12y=66.
- Readmissions with DKA episodes were 3, all above 12y of age, MDI(2) vs IP(1).
- Admissions with severe hypoglycemia were 8,75% on MDI and 25% on IP.
- Based on modality of BG monitoring: best GC noted in group IP using CGM vs SBGM in group MDI. GC was worst in those using FGM in both groups.

Conclusions

- IP usage led to statistically significant improved glycaemic outcomes irrespective of age.

- CGM usage results in improved GC irrespective of whether using IP or MDI.
- FGM was beneficial in only those younger than 12y & offered no advantage in improvement of GC over SBGM in >12y irrespective of modality of Insulin delivery.
- Overall GC was best in <12y irrespective of Insulin Delivery Modality
- Complications of DKA and severe hypoglycemia episodes were more pronounced in those on MDI.

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STUDY OF ASSESSMENT OF CARDIAC AND HEPATIC IRON OVERLOAD IN THALASSEMIA SYNDROME CASES

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Background Chelation therapy has been used to eliminate excess iron. The effective management of thalassemic patients, especially in the paediatric age group, requires optimal monitoring of the toxic effects of both iron overload and excessive chelation therapy. Serum ferritin has been widely used as a surrogate marker and a target ferritin level of 1,000 ng/ml is generally recommended.

Objectives Study of assessment of cardiac and hepatic iron overload in thalassemia syndrome cases

Methods This study was conducted in a tertiary care hospital where thalassemic patients receive regular transfusions. Patients aged 13–33 years coming for routine blood transfusions at the hospital blood bank and also out-patients coming for regular follow up and receiving transfusions at other blood transfusion centres between November 2011 and November 2012 were included in the study. A detailed history and physical examination were completed for all cases and the findings recorded on a proforma.

Results Out of 53 cases the average hepatic iron overload over a study period shows that 34% (18) cases have severe hepatic iron overload, 39.6% (21) cases have moderate hepatic iron overload, 17% (9) cases have mild hepatic iron overload and only 9.4% cases have normal hepatic iron status.

Conclusions Arterial stiffness increases significantly as cardiac iron overload increases but there is no correlation between arterial stiffness and hepatic iron overload. As stiffness index is an indirect measure of CIMT and early atherosclerosis, it also shows and indirect correlation of CIMT with increasing cardiac iron overload. Thus our findings support the hypothesis that iron overload is a risk factor for early atherosclerosis and cardiovascular disease.

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SAFETY AND EFFICACY OF NURSE-LED SALINE ENEMA ADMINISTRATION FOR THE TREATMENT OF MECONIUM OBSTRUCTION OF PREMATURITY

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Background Meconium obstruction of prematurity (MOP) is the third most common cause of neonatal intestinal obstruction, especially in preterm infants. Delayed diagnosis and treatment of the condition can affect morbidity, mortality, hospital stay and the treatment cost of affected preterm infants. MOP can also lead to sub-acute/acute intestinal obstruction