Camptodactyly in CACP is usually bi-lateral & congenital, but in some cases, it develops in early childhood. The degree of contracture need not be equal in both & the deformity may progress or not improve.

Arthropathy principally involves large joint such as elbows, hips, knees and ankles. Histopathologic analysis of synovial tissue reveals pronounced hyperplasia of synovium without evidence of inflammatory cell infiltration or vasculitis, while synovial hyperplasia in rheumatoid arthritis is associated with chronic inflammation.

Non-inflammatory pericarditis is reported in 30% of CACP, this may be mild and self-limited. The presence of coxa vara is noted in 50% of published CACP cases.

Diagnosis is based on clinical findings and confirmed by genetics. Patients with CACP have mutations on the gene PRG4 which encodes the secreted protein called lubricin. Lubricin is a protein that lubricates joints and works as a lubricant between the two layers of the pleura and pericardium.

CACP is often mis-diagnosed as JIA because both present with joint swelling.

At present there is no cure or specific treatment for CACP. Much of the research to date is investigating mechanical benefits of lubricin allowing lubrication of joints. Treatment options are physiotherapy and analgesia focusing on relieving symptoms of the disease.

Conclusions CACP mimics JIA due to similar presentation hence causing a delay in diagnosis & probably unnecessary treatment with anti-rheumatic drugs including biologics. Although it is very rare condition, if there is no or poor response to various immuno-suppression treatment in patients with JIA, we recommend looking into non-inflammatory arthropathy and possible CACP.

British Association for Community Child Health

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ADVANCE CARE PLANS FOR CHILDREN WITH LIFE-LIMITING CONDITIONS ADMITTED TO PAEDIATRIC CRITICAL CARE

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10.1136/archdischild-2021-rcpch.329

Background With advances in medical therapies, and increasing use of long-term ventilation, the number of children living in the UK with life-limiting conditions is increasing. Advance Care Plans (ACPs) allow families and, if appropriate the child themselves, to make important decisions about their care in an unpressurised environment. ACPs are essential to providing quality care for children with life-limiting conditions¹ but are often initiated later than considered optimal.

Objectives I aimed to investigate how many children admitted to Paediatric Critical Care (PCC) with a life-limiting condition already had an ACP in place and for those children without an ACP, if their deterioration could have been pre-empted and ACP discussions started earlier in a more appropriate setting.

In addition, I looked into the accessibility of ACPs on Electronic Patient Records (EPR) to assess the sharing of information between the teams involved in the child's care.

Methods Over a 6 month period, the records of children admitted to PCC with a pre-existing life-limiting condition were reviewed to determine the following:

1. Evidence of an existing ACP
2. Documentation of ACP and resuscitation status on EPR
3. Any ACPs completed during PCC admission
4. Discussions regarding ACP during PCC admission
5. Previous number of PCC admissions

Results 32 patients with a life-limiting condition were admitted to PCC over the 6 month period. Their diagnoses included Rett syndrome, hypoxic-ischaemic encephalopathy and neuro-degenerative disorders. 10 out of 32 children had an ACP in place prior to admission. Three children had ACPs completed during admission, following which one child died after withdrawal of care, and discussions about initiating ACPs occurred in three further patients. Of the patients with ACPs, 70% were for full resuscitation. The number of previous admissions to PCC ranged from zero to ten. Two-thirds of children without an ACP had at least one previous admission to PCC. The ACPs were often difficult to locate within our digital patient records and only two ACPs were recorded on the electronic Carevue system specific to PCC and four on hospital-wide EPR.

Conclusions The majority of children with ACPs were for full resuscitation, and all were admitted to PCC, highlighting that not all ACPs equate to ceilings of care or 'do not resuscitate' decisions, but can instead be used to express the family's wishes for their child's care. Overall 20% of families were involved in ACP discussions during their child's admission to PCC. This is a stressful and time-pressured environment in which to expect families to make such important decisions. We need to empower general and community paediatricians to start ACP discussions with families earlier, which would allow families the time and space to make these decisions. In addition, earlier implementation of ACPs can serve as a platform for further discussions in the event of the child being admitted to PCC acutely unwell. However, for ACPs to be fully effective we need a facility to allow ACPs to be easily located on EPR in order to facilitate sharing of information between teams involved in the child's care.

REFERENCE

1. The End-of-Life Care Strategy, Department of Health, 2008.

British Society of Paediatric Endocrinology and Diabetes

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STUDY OF VISFATIN AND FETUIN-A IN TYPE 2 DIABETES MELLITUS IN CHILDREN

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10.1136/archdischild-2021-rcpch.330

Background T2DM is a progressive disease with which can affect multiple body systems. And has a rapidly increasing prevalence.

Visfatin is one of the adipokines which might play a role in the pathogenesis of T2DM, insulin resistance and all parameters of metabolic syndrome.

Also Fetuin-A is a protein synthesized by the liver and released in the circulation. It resembles serum albumin but is more abundant during fetal life. It is involved in several functions such as endocytosis, brain development and the formation of bone tissue, however its exact significance is still obscure, its importance in T2DM, Insulin resistance is still debatable.

The aim of this study is to assess the level of fetuin-A and visfatin in patients with T2DM and to correlate their levels with clinical and other biochemical variables in patients

Objectives Assessment of the level of novel markers fetuin-A and visfatin in patients with type 2 diabetes mellitus and to correlate their levels with clinical and other biochemical variables in such patient

Methods This case control study was conducted on 88 participants divided into two groups.

First group included 44 already diagnosed type 2 diabetes (cases), 19 males and 25 females, their age ranged between 10 and 16 years. They were selected by stratified random method from outpatient clinics and inpatients of Mansoura children hospital.

Second group included 44 apparently healthy non diabetic individuals of matched age and sex (control group). Their non-diabetic state was confirmed by oral glucose tolerance test. A written consent was obtained from all participants.

Both groups were subdivided into 2 subgroups, obese group ($\text{BMI} \geq 30 \text{ kg/m}^2$), non-obese group ($\text{BMI} < 30 \text{ kg/m}^2$)

Results There was no significant difference between all study groups as regard age, gender, smoking, systolic, diastolic, mean blood pressure, LDL-C and total cholesterol (all p -value > 0.05).

There was a significant difference between all study groups as regard body mass index, HDL-C, TG, fasting blood glucose, 2 hour post-prandial blood glucose, fasting insulin, HOMA-IR, HbA1c, visfatin and fetuin-A (all p value < 0.05)

Conclusions Our study concluded that serum visfatin levels were higher in patients with T2DM versus control subjects also serum visfatin was high in obese subjects versus non obese whether diabetic or non-diabetic and there was positive correlation between visfatin level and HOMA-IR, FPG and BMI.

The increased level of serum visfatin in T2DM may be related to obesity, hyperglycemia which could induce visfatin release or it is a compensatory mechanism to ameliorate insulin deficiency with progressive B cell dysfunction in insulin resistant patient, or finally due to adipose tissue inflammation in insulin resistant subjects.

As regard Fetuin-A we showed that its level is deficient in T2DM and also decreased levels had been decreased in obese versus non obese subjects with negative correlation with HOMA-IR, BMI and FPG which may be due to some medications used like metformin and pioglitazone which were established to reduce the level of fetuin-A or due to non-enzymatic glycation which mask the effect of fetuin-A on insulin resistance due to glucose toxicity.

Quality Improvement and Patient Safety

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TO BLEEP OR NOT TO BLEEP? IMPROVING NIGHTSHIFT WORKING FOR PAEDIATRIC JUNIOR DOCTORS

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10.1136/archdischild-2021-rcpch.331

Background Healthy nightshift working for doctors' wellbeing and patient safety is well documented. However anecdotally many tier-1 trainees (junior residents) struggle to take natural breaks due to non-urgent tasks communicated via the 'bleep' system.

Objectives Our aim was to record bleeps made to the tier-1 trainee over 7 nights to investigate whether there were opportunities to improve nightshift working.

Methods The study ran over 7 consecutive nightshifts (20:30–08:30) in November 2019 at a large tertiary children's department. One tier-1 trainee is supported by one tier-2 trainee (senior resident) overnight. All bleeps were logged over the period described. We analysed the bleep frequency, reason and timing.

Results 173 bleeps were received over 7 nightshifts with a mean (SD) of 24.7 (7.1) bleeps/night and 2.1 (1.6) bleeps/hour. 33 bleeps (19.1%) were for urgent patient reviews. 40 (23.1%) were prescription queries. 11 (6.4%) to re-site cannulas. 18 (10.4%) were laboratory result reviews. 19 (11%) were made in error. Only 5 bleeps (2.9%) were for multiple queries. Between 2:30am–5:30am, the natural circadian nadir, there were 6.6 bleeps (SD 1.4); 64% were scored as non-clinically urgent.

Conclusions

- We found trainees are bleeped approximately once every 30 minutes between 2:30am–5:30am, nearly two-thirds of which could have waited.
- Non-urgent tasks should be limited out-of-hours and trainees encouraged to take natural breaks.
- This could be facilitated by bleeps between 2:30am–5:30am being escalated via the ward nurse-in-charge.
- Bleep frequency tended to peak after the nursing handovers, often coinciding with doctors' handovers. Distraction during this period may lead to doctors missing important clinical information.
- Altering daytime prescribing practise to cover for the night time could reduce the prescribing pressure for a doctor over night.
- We advise offering guidance to nursing teams to cohort communication of non-urgent tasks; minimising incorrectly directed bleeps and interruptions to clinical reviews.

Quality Improvement and Patient Safety

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MANAGEMENT OF CROUP: A QUALITY IMPROVEMENT PROJECT

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10.1136/archdischild-2021-rcpch.332