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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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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