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Review

Critical timing of gastrostomy insertion in a child with cystic fibrosis

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SUMMARY

Pulmonary exacerbations and malabsorption in children with cystic fibrosis (CF) can lead to faltering growth and poor weight gain. Children with a higher BMI (body mass index) show a slower decline in lung function. Our specialist CF centre has experienced a death following gastrostomy insertion in a young CF child, despite maximal medical intervention, which has made us reflect on our practice and the urgency with which we discuss the option for a gastrostomy to improve nutrition.

INTRODUCTION

Cystic Fibrosis (CF) is a characterised by recurrent pulmonary infections and chest exacerbations. A significant proportion of patients have pancreatic insufficiency leading to intestinal malabsorption. The combination of recurrent infection and malabsorption often leads to poor weight gain and growth. The link between nutrition and pulmonary health in CF is well established. Improvements in weight gain and body mass index (BMI) are associated with a slower decline in lung function [1–3], and malnourished patients show a faster rate of annual lung function reduction [4]. In malnourished children and adolescents, lean body mass is also strongly associated with lung function [5]. In our practice, we target patients who fail to regain birth weight, drift across weight centiles, or have a BMI $<25^{th}$ centile (www.rbht.nhs.uk/childrencf).

The first step in achieving optimal BMI or weight involves dietary advice and monitoring, hence the importance of specialist paediatric dietetics in a CF multi-disciplinary team. The next step is assessment for causes of poor weight gain, such as inadequate or low adherence to pancreatic enzyme replacement therapy, impaired glucose metabolism or overt CF-related diabetes, low urinary sodium and coeliac disease. Once everything possible is corrected, the use of oral calorie supplementation is introduced, although a Cochrane systematic review showed no additional benefit above dietary advice and monitoring alone in moderately malnourished children with CF [6]. If improvement is not seen, the option of direct gastric feeding with the insertion of a nasogastric tube or gastrostomy is discussed. Criteria to introduce gastrostomy feeding between CF centres is not fixed, however in this patient group, a gastrostomy has been shown to increase the potential of a child achieving a BMI above the 50^{th} percentile as advised by the North American CF Foundation [7] and improve other anthropometic markers of nutrition[8–10].

Data from our centre showed improvement in weight and BMI following gastrostomy insertion but no improvement in lung function. We inserted 77 gastrostomies in a 20 year period from 1990-2010. Weight and BMI significantly increased in the 12 months following gastrostomy: weight z score +0.6 ($p<0.001$) and BMI z score +0.74 ($p<0.01$). Forced expiratory volume in 1 sec (FEV_1) z score measurements did not significantly change during the 12 months following gastrostomy insertion [11].

Patients and families are often hesitant about a gastrostomy insertion, as it is perceived to be invasive and can affect the child's body image. Counselling by the CF team is required, and if prolonged will delay the procedure. Once the tube is inserted and in regular use, the overall satisfaction with the system is generally high with many carers stating they wished they had instigated the procedure earlier [12]. Parent and child relations and the often fraught mealtime experience can also improve hugely [13].

CASE HISTORY

The patient was a six-year-old girl, diagnosed on newborn screening at 3 weeks of age, with the classic genotype homozygous p.phe508del. She was pancreatic insufficient and was diagnosed with CF-related liver disease aged 4 years. She frequently grew *Burkholderia cepacia* and *multivorans* in cough swabs and sputum samples, despite attempts at eradication, and the first isolation of

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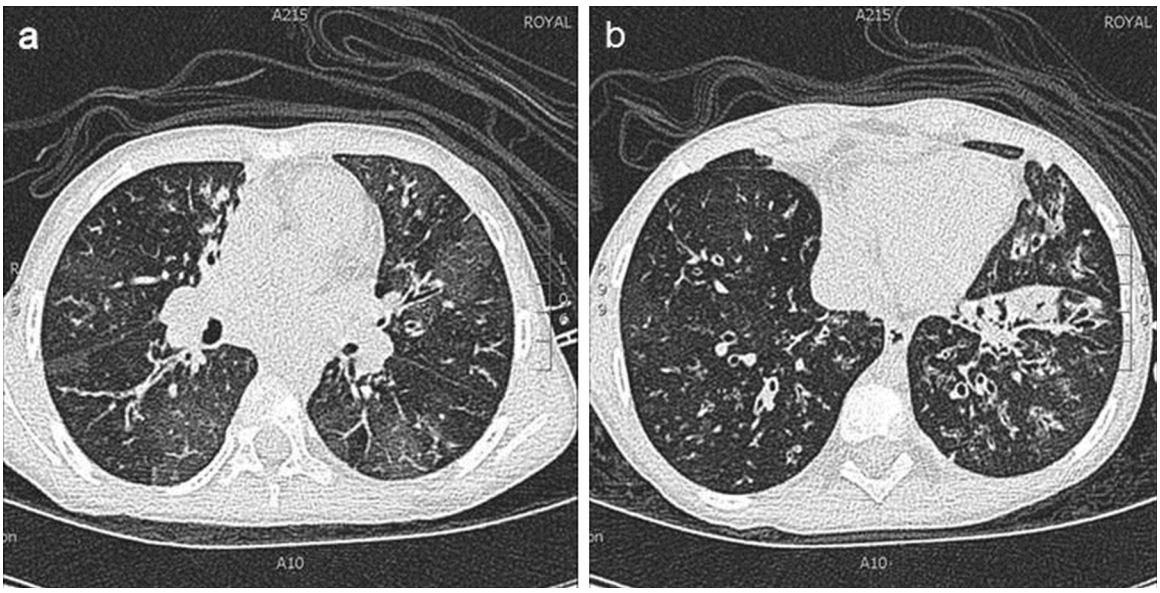

Figure 1. Chest CT images from a scan performed 9 months before the gastrostomy insertion, showing (a) mosaicism suggestive of severe small airways disease, and (b) dilated airways consistent with bronchiectasis.

Burkholderia when she was just 3 years old. She was admitted for routine intravenous antibiotics every 3 months, to maintain lung health, and she was seen frequently between admissions in clinics and at home.

Over a two-year period her lung function dropped precipitously from an FEV_1 of 80% predicted in autumn 2012 to 40% predicted by spring 2014. She was receiving a significant degree of home therapies between admission in the form of 7% hypertonic saline, dornase alfa, rotating nebulised antibiotics (colistin and tobramycin), inhaled corticosteroids, and oral theophylline treating both bronchiectasis and severe small airways disease (Figure 1). Over the year, it was noted that when she came into hospital, her presentation was increasingly severe, so she frequently required supplemental oxygen and on one occasion, non-invasive ventilation (NIV) in the form of BiPAP (bilevel positive airway pressure) at the start of the admission.

There had been concern regarding her weight by the CF team for some time, as her weight had fallen to below the -2 z-score. This was despite oral calorie supplementation with Enshakes and calogen-based drinks. The subject of gastrostomy feeding was introduced in the winter of 2013 when no improvement in weight had been seen. The decision to opt for gastrostomy insertion took considerable effort and counselling on our part and was agreed after 7 months of discussions. Interim nasogastric feeding had been considered however, both parents and the CF team felt this would not be tolerated and she would pull the tube out immediately.

She was admitted pre-operatively for two weeks of intravenous antibiotics and intensive physiotherapy. At this point her FEV_1 was 40% predicted and her mean oxygen saturations overnight were 95%. After induction of general anaesthesia, thick airway secretions made oxygenation difficult, and so bronchoscopic lavage was undertaken to aid airway clearance. The endoscopic gastrostomy insertion was uneventful. She was admitted to the paediatric intensive care unit post-operatively as planned and extubated within 4 hours into supplemental oxygen, and remained stable throughout the first night.

Ten hours later however, she developed acute respiratory distress and a chest radiograph showed a right-sided tension pneumothorax requiring an urgent chest drain. Her respiratory support escalated from NIV, to intubation with conventional ventilation, and eventually high frequency oscillation ventilation (HFOV) over a six-day period. A trial of lung lavage was instigated with the child on arterio-venous Extracorporeal Membrane Oxygenation (ECMO) support. This option was decided upon as her deterioration was post-operative, rather than part of a gradual decline in an end of life situation, so it was hoped it would be reversible, particularly in light of her young age. Two litres of 0.9% saline was used to lavage both lungs three times over a period of 48 hours however, this intervention led to no improvement and sadly the situation was then considered futile. After multiple discussions with her parents, care was withdrawn and she died peacefully.

DISCUSSION

Death in childhood has become uncommon in the modern era of CF. All deaths in CF children at Royal Brompton Hospital and Great Ormond Street Hospital over a ten-year period were reviewed and published [14]. There were eleven deaths from 2000-2009 with a female preponderance. Nutritional status was generally poor and 68% had a gastrostomy. The majority died of respiratory failure (64%) and were ventilated (45%). Care was withdrawn on six of the eleven children. Only one died of a postoperative complication, a cerebral haemorrhage following bronchial artery embolization for haemoptysis on the background of an unexpected anomalous artery circulation. Our recent experience adds to this published series confirming that death in paediatric CF is often unpredictable, thus active management continues throughout and often occurs in hospital involving ventilation. Instigating formal palliative care may be difficult in these situations.

The use of ECMO in CF is a controversial subject owing to the progressive nature of the disorder. A recent series from the Extracorporeal Life Support Organization (ELSO) identified 77 ECMO episodes used on adult CF patients with an overall survival rate of 52% [15]. Owing to the increasing use of ventilation for acute exacerbations in CF, a prospective study evaluating outcomes following ECMO has been suggested. To our knowledge, whole lung lavage has not been reported in CF previously. This was the first attempt at our centre in a child with CF, however we have substantial positive experience in its use for pulmonary alveolar proteinosis.

Our experience with this child has raised several question surrounding procedures requiring general anaesthesia and the timing of gastrostomy insertion patients with severe lung disease. Like many centres, we have dedicated paediatric anaesthetic support with great experience in ventilating children with critical heart and lung disease. We will often opt for a general anaesthetic for relatively minor procedures such as obtaining venous access in difficult cases. The risks associated should not be underestimated. We will also often undertake an opportunistic bronchoscopy for microbiology samples from the lower airways in non-sputum producing children. This will inevitably add additional strain to the ventilation of these children during and after the procedure, although in the case discussed here, the aim of the bronchoscopy was to clear secretions and aid ventilation not to sample lower airway secretions.

The most pressing question is whether we stress sufficiently to parents the potential risks of delaying procedures aimed at improving weight gain and therefore lung function. If discussions surrounding gastrostomy insertion are instigated early, the child may not only be in a healthier state to undergo the procedure, but also avoid unnecessary decline in lung function. However, when faced with parent resistance to a procedure we believe to be necessary, should further steps be taken to force the issue involving social services intervention and legal involvement? This may benefit the child in the long run but is bound to affect the working relationship with the family, which would be detrimental.

CONFLICT OF INTEREST

None

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