In recent years the numbers of children with complex and continuing health care needs have been increasing within the United Kingdom (Kirk, 2008; Hobson and Noyes, 2011). Whilst a relatively small group, these children and their families require a high level of care and support from a wide range of professionals, services and organisations, to ensure their needs are met as much as possible in their own home (Ward et al., 2014).

Liam is a ten month old boy who was diagnosed as having Cardiac-faciocutaneous (CFC) syndrome at birth. As a direct result of his condition he has ongoing complex health care needs. Liam lives with his parents, Alan and Cora, in a 2 bedroom house. Liam was a much longed for first child. After Liam’s birth and his diagnosis, Alan and Cora experienced both emotional and practical chaos (Price et al., 2011). Alan works away a lot and Cora (who gave up her job after Liam was diagnosed) carries out all Liam’s care with input from the community children’s nursing team. Close links are maintained with the Paediatrician at the local hospital.

CFC syndrome is a rare genetic condition (present at birth), typically characterised by its effect on the heart (cardio), facial features (facio), skin (cutaneous), resultant learning disability, and feeding difficulties in early life (Contact a family, 2014). As one of the RASopathies CFC bears many clinical features in common with the other syndromes in this group, most notably Noonan syndrome and Costello syndrome (Pierpoint et al., 2014). CFC is classed as an autosomal dominant condition, which means one copy of an altered gene in each cell can lead to the disorder. CFC usually results from new gene mutations and occurs in people with no history of the disorder in their family. Due to the multi-pathological nature of CFC syndrome, this paper focuses on Liam’s feeding problems, particularly gastroesophageal reflux (GER) and faltering growth, alongside the new recently experienced issue of seizures.

Schwartz (2014) state that GER is caused by immature function of the lower oesophageal sphincter, which frequently relaxes, allowing an involuntary retrograde flow of gastric contents into the oesophagus (Mersch, 2014). As a consequence of the increased acid the child can experience persistent vomiting after feeding as well as discomfort in their mouth and throat. Resultant problems are therefore often directly related to poor weight gain and faltering growth (Czinn & Blanchard, 2013). Such problems were evident with Liam as he had not been successfully bottle feeding and had a history of vomiting - subsequently his weight had decreased. A Percutaneous Endoscopic Gastrostomy (PEG) had previously been inserted, essentially bypassing the oesophagus, and feeding directly into the stomach (Marks, 2015). Seizure activity is also a common symptom in CFC syndrome (Armour & Allanson, 2008), caused by a possible delay or absence of myelination in the neural pathways (Aizaki et al...
2011). Liam had recently been experiencing seizures.

While decision making is continuous within the planning and provision, the focus here is on a day when both the community nurse and health visitor visited the family home. Providing quality care and addressing the needs of Liam’s care - something that parents also highly anxious about seeing Liam having enteral feeding has positive effects on children with complex needs who experience feeding problems for children with complex health care needs. Parents appear to take on the role of parents who care for children with complex needs, hence Standing (2009) with findings indicating that many parents correlate the child's care with the role they often feel in supporting. Such a belief was explored in a qualitative study with 47 parents of children with complex health care needs. It is important to be cognisant of the fact that children with complex needs often have very large and varied teams of professionals working together and that the members of the team may change in tandem with the child's condition. A range of professionals working together can pose problems, hence Standing (2014) asserts that collaborative decision making is particularly important for those with complex health needs. Parents are central to team functioning, with Smith et al. (2013) stating that expert parents in the family can be invaluable in maintaining the child's care - a role they often feel in supporting. The evidence based model proposes that they be SMART: specific, measurable, achievable/agreed, realistic and time - limited. Since Liam’s recent failure to gain weight initially prompted the home visit (he had dropped a weight centile due to insufficient bottle feeding), his weight loss and reduced nutritional status was the focus and main goal of care. The first goal set was specifically regarding the aim of promoting Liam’s nutritional status, to be measured through a desired increase in his weight at the next visit in two weeks. In addition, Liam had experienced a new problem, seizures. The goal here was to maintain Liam’s safety during seizures, something also to be reviewed at the next visit. The challenge of increasing feeding tolerance and increasing anxiety and isolation stemming from both the feeding difficulties and from fearing eating during the seizure, a third goal was set. This was a longer term goal and aimed to reduce Cora’s anxiety and isolation through information and support.

Given the goals set, decisions about the plan of care therefore centered around managing his feeding and associated weight loss, managing his seizures and addressing associated maternal anxiety. The use of best evidence was emphasized, particularly regarding management of Liam’s GER and seizures. The evidence based model of decision making originally identified by Sackett (2000) involves a tripartite approach which includes best available research evidence, professional expertise and patient preferences/needs. Decision making does not solely incorporate research evidence but is often based on a combination of evi...
References


