Unresolved grief in parents of children with CF: A pilot randomised controlled trial on the use and delivery sequence of disease-related education and psychotherapeutic support

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ONLINE RESOURCE 1

A) OUTLINE OF BASIC PARENTAL EDUCATION ABOUT CYSTIC FIBROSIS

- What is cystic fibrosis?
- Basic genetics
- Details about organs affected
- Essentials about management options, with focus on the respiratory system
- Overview of support provided by hospital
- Overview of support provided by community
- Prognosis and what to expect
- Time for questions

B) OUTLINE OF GASTROENTEROLOGY EDUCATION ABOUT CYSTIC FIBROSIS

- Interactive, simple, visual based educational slides in a non-rushed setting.
- Time allocated 45 minutes, while prepared material is delivered through power point but more emphasis is given on answering parent's questions. Following issues are covered.
    - How does a normal gastrointestinal system work?
    - How is gastrointestinal system affected in cystic fibrosis?
    - Need for nutritional surveillance?
    - What to expect in GI CF clinic appointments?
    - Overview of support provided by specialist gastroenterologist?

- Time for questions

C) OUTLINE OF NUTRITIONAL EDUCATION ABOUT CYSTIC FIBROSIS

Overview of nutritional aspects of CF

Good nutritional status in Cystic Fibrosis is imperative to achieve normal growth and development. As a result of the pathophysiology of cystic fibrosis, there are numerous barriers to achieving optimal nutrition. The prevalence of malnutrition in CF is common and generally results from a combination of reduced oral intake, increased energy expenditure and fat malabsorption which results in negative energy balance and weight loss/failure to gain optimal weight.

Common barriers to achieving optimal nutritional status in CF include the following;

- Increased energy expenditure
- Deranged bowel actions
    - Nutrient malabsorption
    - Cramping, abdominal pain, abdominal distension, constipation, steatorrhoea
- Poor oral intake
    - Poor appetite, early satiety, fussy eating
    - Illness & infection (raised inflammatory markers related to anorexia)
- Frequent coughing resulting in nausea and vomiting
- Additional conditions of CF
    - Reflux (GOR)
    - CF-related diabetes (CFRD)

    - Distal intestinal obstruction syndrome (DIOS)
    - CF-related liver disease (CFLD)
    - CF-related osteoporosis
    - Anaemia of chronic disease
- Behavioural/psychological problems

The role of the dietitian

- To achieve normal growth and development
- To optimise absorption of nutrients by appropriately using Pancreatic Enzyme Replacement Therapy (PERT)
- To replace electrolytes lost through increased sweat levels.
- To educate patients and their parents regarding other factors associated with CF (e.g. fat soluble vitamin deficiency, altered gastric motility, impaired glucose tolerance / diabetes, and reduced bone mineral density)

CF diet

- High energy, high salt
- Shift from high fat to high energy (including healthy fats and high protein)

The importance of PERT

- What is PERT?
- How to use it/how to administer it
- Fat counting and dosing including resources to support this process e.g. books, phone apps

Why is salt important?

- Recap of pathophysiology of CF and increased salt losses
- How to use it/how to administer e.g. add salt to food, salt liquid, salt tablets, electrolyte/sports drinks
- Winter vs Summer dosing
- Increased requirements with exercise and activity
- Signs and symptoms of inadequate salt

Fat-soluble vitamins

- Reiterate pathology of CF in the context of fat malabsorption
- How to use it/how to administer e.g. crushed, swallow whole, syringe
- Prescription for age

D) OUTLINE OF BASIC PHYSIOTHERAPY CARE CYSTIC FIBROSIS

Why is physiotherapy needed for children with cystic fibrosis?

Chest physiotherapy treatment is able to:

- Clear Mucus from the lungs to help prevent chest infections
- Remove excess mucus when an infection is present
- Help the lungs work normally

Chest Physiotherapy needs to:

- Be ongoing even when no symptoms are obvious
- Start as soon as possible after diagnosis of cystic fibrosis

How do the lungs work?

- The lung
- The airways
- The alveoli
- The muscles of breathing

What is involved in chest physiotherapy?

- Mechanism of clearance
- Nebulised Solutions
- Positioning
- Technique
- FET
- Exercise and activity

How may chest infections be identified?

- Cough
- Mucus
- Changes in breathing.
- Increased tiredness or reduced activity
- Decreased appetite and/or weight loss
- Increases irritability
- Fever

How will physiotherapy treatments fit into our daily routine?

How will Physio change as my child grow