Children with Cystic Fibrosis

What is Cystic Fibrosis?

Cystic fibrosis (CF) is a hereditary disease of the *mucus, saliva and sweat glands. It affects organs such as lungs, pancreas, liver, intestines, sinuses, and sex organs* (MedicineNet Inc, 2009).

"Cystic fibrosis is the most common inherited, life limiting, incurable condition amongst Caucasians" (Royal Children's Hospital)

What Causes CF?

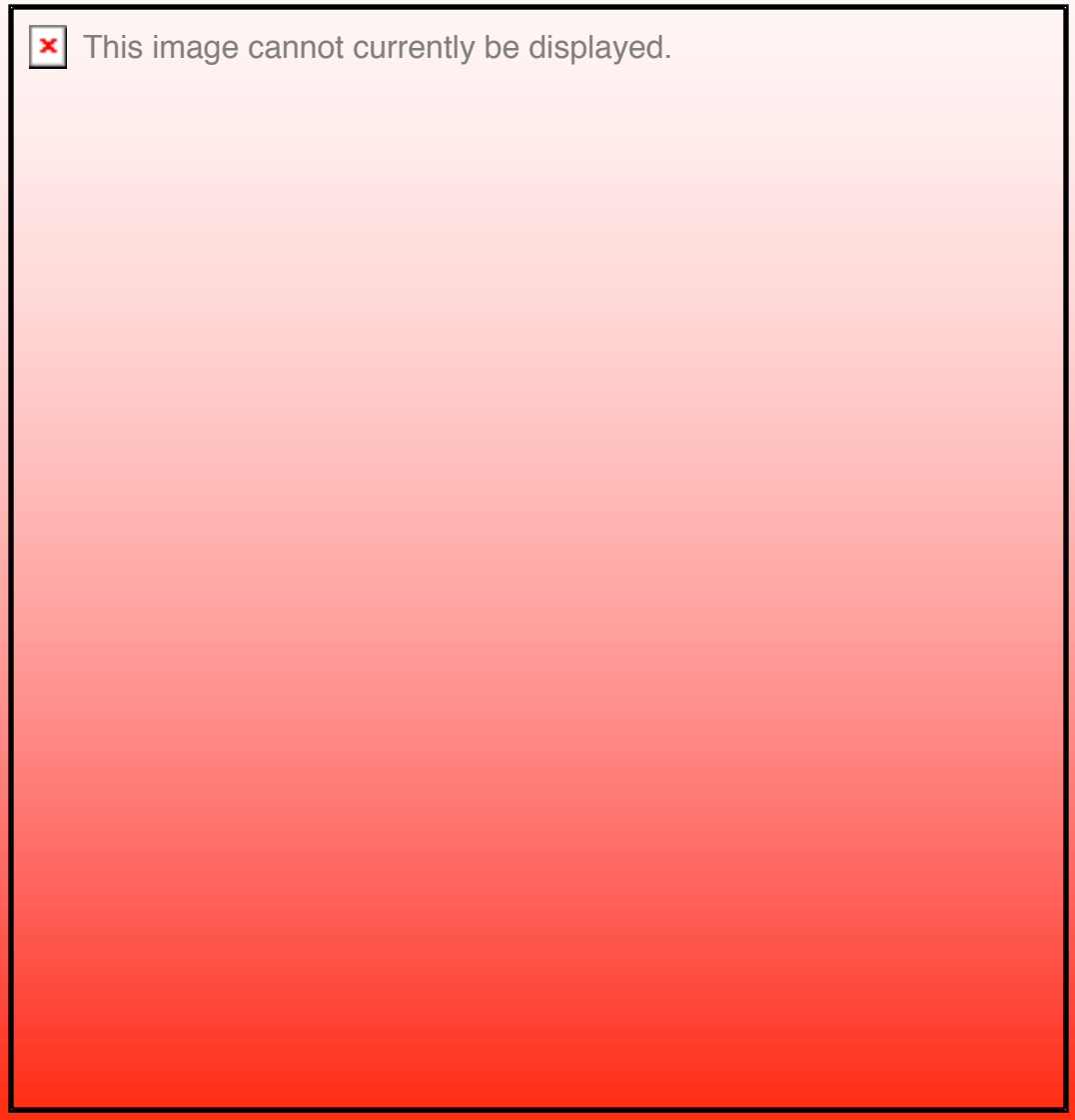
• Cystic Fibrosis is an inherited disease.

• For a child to inherit CF, both parents must be carriers of a defective gene on chromosome 7.

- They then have a 50% chance of becoming a carrier.

- A 25% chance of getting CF

- A 25% chance of not being a carrier and not having CF



What is it about Chromosome 7 that causes CF?

- A chromosome carries genetic information
- Chromosome 7 carries the cystic fibrosis transmembrane conductance regulator (CFTR)
- CFTR controls salt and water movements in and out of cells
- When CFTR is defective, cystic fibrosis occurs because the CFTR doesn't work or is completely missing.
- When salt and water don't move in and out of cells properly, sweat becomes 5 times saltier and a thick, sticky mucus is produced outside the cell.

What does the thick, sticky mucus do?

It affects the…

Lungs

- Mucus builds up and obstructs airways
- Build up also makes a suitable environment for bacterial growth

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Bacterial growth increases risk of infections

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Repeated infections cause lung damage

Pancreas

- Pancreas produces enzymes that help with digestion
- Build up of mucus blocks ducts in pancreas, stopping enzymes form reaching intestines

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Without enzymes, intestines can't digest food properly

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Leads to loss of vitamins and nutrients

Symptoms of CF

- Persistent cough, often with phlegm or possibly blood streaking
- Difficulty breathing / wheezing
- Fatigue
- Fleshy growths inside the nose (nasal polyps)
- Weight loss / poor weight gain
- Frequent lung infections, pneumonia, bronchitis
- Abdominal pain
- Salty tasting skin (salt loss when sweating) leading to dehydration
- Greasy, light coloured, foul smelling stools or diarrhoea

Implications For Development: Social / Emotional

- Child may have difficulty interacting with other children due to periods of absence from centre
- Child may experience some emotional distress – depression, withdrawal, becoming over dependent
- Child may feel embarrassed by effects of their CF. For example – coughing a lot and bringing up phlegm.

Implications For Development: Physical

- Children with CF are very capable of participating in normal physically active play and activities.
- Regular exercise is important – it helps to loosen mucus
- Child's current health status may not allow them to fully participate in physical activities. They can still participate but may need more frequent breaks to cough and have a drink.

Implications For Development: Cognitive

- A child's cognitive development is not usually affected by cystic fibrosis
- Malnutrition can stunt cognitive development. Research conducted by The American Academy of Pediatrics suggests children with CF are at risk of malnutrition because of their loss of vitamins and nutrients. This is mainly a concern when CF hasn't yet been diagnosed.

Implications For Development: Language

- A child's language development is not usually affected by cystic fibrosis.
- If the child is sick in bed at home or the hospital, reading stories to them is a great opportunity to promote early language and literacy skills.

Modifications to the Service

- No structural modifications are usually required.
- There are a few modifications needed to be made to the program to ensure the health and safety of the child with CF, mainly during routine times.
- Resources to assist the child while at the centre may include their nebuliser. Tissues are also helpful to reduce embarrassment when coughing up phlegm.

Involving Family in the Modifications

- Involving a child's family in the service and maintaining effective communication is vital in developing an individualised routine for a child with CF.
- The family will be able to tell you about their child's cystic fibrosis and how it effects their child - Just as every child is different, each child with cystic fibrosis is also different!!

Individual Routines - Physiotherapy

- Parents may need to come to the centre to assist the child with their daily physiotherapy.
- Physiotherapy helps clear the airways of the mucus in the lungs and is important in helping to reduce the risk of infections.
- The amount of physiotherapy sessions each day varies with each child and each day.
- There are a number of physiotherapy techniques a child with CF may use, some include: chest percussion, vibrations and breathing techniques.

Individual Routines - Mealtimes

• Children with CF need to eat extra calories and protein than other children. This is so they can replace the fat and protein they pass and be well nourished to assist in preventing infections.

• Children with CF also require supplements at mealtimes. Supplements may include:

- Vitamins to replace vitamins lost in digestion

- Salt supplements, especially in hot weather or when exercising to replace salt lost through sweat

- Pancreatic enzymes to assist with digestion

Reference List

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