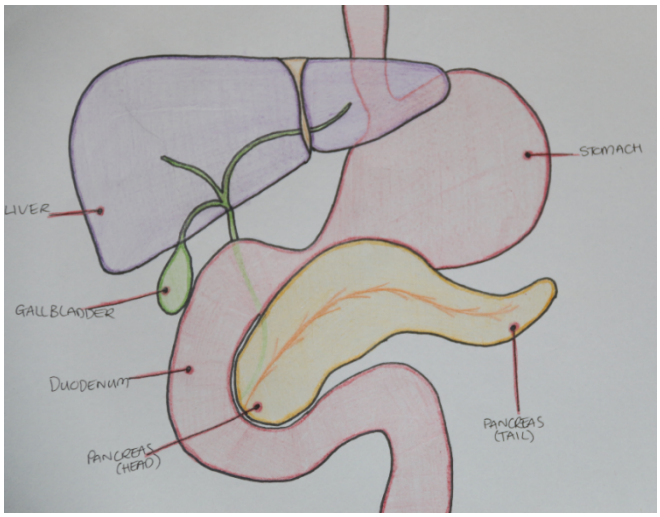
How CF Affects the Gastrointestinal System



Approximately 85% of newly diagnosed babies with CF are unable to fully digest their milk due to problems with their pancreas. The pancreas is an organ that sits behind the stomach, which releases enzymes to assist with the breakdown and digestion of food and milk in the small intestine. In children with CF the pancreatic ducts are blocked reducing the release of digestive enzymes, causing pancreatic insufficiency. If their food and milk cannot be digested and absorbed is called malabsorption. Malabsorption results in poor weight gain or weight loss and loose frequent bowel actions which contain undigested fat. Other symptoms of malabsorption can include bloating and excessive flatus, abdominal pain and cramping.

How to manage pancreatic insufficiency

The diagnosis of pancreatic insufficiency is made through the examination and testing of a stool sample. Pancreatic insufficiency can be treated by giving your baby or child a pancreatic enzyme extract before every feed or meal. We typically refer to these extracts as ‘enzymes’.

Enzymes

Enzymes (eg Creon, Panzytrat) are designed to replace the body’s own enzymes that cannot pass through the pancreatic ducts due to blockage. The enzymes are derived from pig pancreas. They are taken orally as granules in apple puree for babies and capsules for older children. The enzymes will pass through into the stomach and intestines where they help to break down food for absorption. They are specially coated to resist the stomach acid environment but are released from this coating in the small intestine where they will work to digest the food. You will need to give your baby their dose of enzymes before each feed and to your child before each meal and snack to ensure normal digestion.

Your specific enzyme plan will be discussed with your dietician.

How to monitor pancreatic insufficiency and malabsorption

Weight monitoring

Your baby's weight will be closely monitored to ensure adequate nutrition and enzyme replacement is achieved. You will be seen by your maternal and child health nurse. Monash Children's at Home may also do home visits for weight monitoring to liaise with your doctors and dietician as well as weight monitoring at outpatient appointments.

Stool samples

We test stool samples for the amount of fat which will indicate the degree of malabsorption. A high amount of fat in the stool of a baby with CF indicates malabsorption due to pancreatic insufficiency. Taking a stool sample from your baby's nappy is a simple noninvasive test and will continue to determine if enzyme therapy is adequate.

Diet and nutrition for babies

Each baby will be affected by CF in different ways. It is important to remember that each baby with CF will have an individualized nutrition plan. A balanced diet with enough calories and the right amount of mineral and vitamin supplements is the key to health and good nutrition.

Milk

In most cases breast milk or formula is adequate for babies during the first six months. In cystic fibrosis it is important to obtain enough nutrients to allow your child to grow and develop to the best of their ability. Children with CF have a higher energy requirement and increased calories may be needed as directed by your CF dietician.

In babies and children with CF, maintaining adequate nutrition for growth and development can be challenging as a child with CF needs a higher energy consumption. If your child is unable meet their energy requirements through food and fluid orally, they may suffer from

malnutrition and weight loss. A Nasogastric Tube (NGT) or Percutaneous endoscopic gastronomy tube (PEG tube) may be required to assist with your child's energy needs.

NGT

A nasogastric tube may be required to help deliver medications and extra calories or fluid to your baby. It is a soft tube that is inserted into your baby's nose, down their oesophagus and into their stomach. It is inserted on the ward with the nursing staff and we will teach you how to use it if you're required to go home with it in. the NGT will need to be replaced once a month either with a nurse at the hospital or we can teach a parent to insert it at home. The NGT is taped to the child cheek and will be visible.

PEG Tube

Some children need to have a PEG tube surgically inserted. It is a soft silicone feeding tube that is surgically inserted into the stomach. It allows extra fluid and calories to be given directly into the stomach. Although this procedure is reversible it is a longer term alternative compared to a NGT, which may be used prior to making the decision for a PEG tube. It can also be used to deliver medications and can be hidden under clothes.

Solids

Introduce solids to your baby from 4-6 months of age. Be aware that you may need to increase their enzyme dose. Speak with your dietician for more information.

Salt

People with CF including babies lose more salt through their sweat and require extra salt in their diet. Salt supplements are prescribed and for infants it can be taken in a liquid form via a syringe or added to their feed.

Vitamins

Babies with CF often have vitamin deficiencies in fat soluble vitamins A, D, E and K. There is a CF specific multi-vitamin supplement called VitABDECK that is commonly prescribed that can be crushed and mixed with puree or diluted in water to give to babies.

For more diet and nutrition information for babies please visit
www.cfwa.org.au/wp-content/uploads/2017/12/Nutrition-for-Infants.pdf

Notes

References

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More Please: A Guide to Eating for Children with Cystic Fibrosis. (n.d)

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