When and why should my child have a gastrostomy?

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AIMS

By the end of this session participants should be able to:

- Describe the factors responsible growth failure in Ataxia Telangiectasia (A-T), a disease associated with high early mortality from lung disease
- Identify the trajectory of progressive growth failure and pulmonary aspiration in A-T
- Formulate plans to manage growth failure

SUMMARY

Ataxia Telangiectasia (A-T) is a rare autosomal recessive, caused by mutations in the ATM (Ataxia-Telangiectasia mutated) gene. This gene encodes a protein kinase, ATM, which is responsible for the cellular response to double stranded DNA breaks. A-T causes progressive ataxia, immunodeficiency, sinopulmonary infections, oculocutaneous telangiectasia, increased cancer risk and increased sensitivity to therapeutic doses of ionising radiation. The median survival is 25 year; the leading causes of premature death being respiratory diseases and cancer.

Undernutrition adversely affects lung health. Poor nutritional status and decreased pulmonary function have been shown to be linked in other diseases, including Cystic Fibrosis (CF). Worsening nutritional status increases infection-related morbidity and mortality. Malnutrition is of particular concern in children since it adversely affects not only normal accrual of height and weight but may also impact lung development.

Small cross-sectional studies have shown that patients with A-T exhibit high rates of malnutrition, short stature and reduced lean body mass. Numerous factors including neurodegeneration, limited food intake with progressive disease, dysphagia and/or swallowing incoordination, limited physical activity, hormonal changes, hypogonadism, insulin resistance, glucose intolerance, abnormal expression of IGF1 (somatomedin C), low levels of IGFBP3 (insulin-like growth factor binding protein-3), and infections and an associated hyper-catabolic state all potentially contribute to poor growth. A-T causes extreme insulin resistance, but clinical diabetes is diagnosed infrequently and it is unclear whether the presence of diabetes also affects nutritional status and lung disease. Oropharyngeal dysphagia with aspiration is common and is progressive in older patients (second decade) with A-T. The onset of dysphagia coincides with a decrease in nutritional status, although in a cross sectional study it is not possible to distinguish between nutritional deficiency as a cause or effect of dysphagia. Hence, it is important to maintain good nutrition in children with A-T to protect respiratory function and to ensure normal growth.

There is a clinically important and statistically significant decline in weight and height Z-scores over time. Despite regular dietician advice, fortification of food and prescription of nutritional supplements, approximately 1/4 of A-T patients are underweight and/or stunted (Z-scores less than -2). The drop in weight Z-score appears to commence at around the 8th birthday. 70% of these children improved their weight Z-score after institution of gastrostomy feeding.
Aspiration can cause lung disease in children with neurological disease, can be silent and can lead to bronchiectasis early. Assessing for presence of aspiration can be challenging especially in A-T because of concerns regarding radiation exposure. As well as growth failure, presence of aspiration is another indication for gastrostomy insertion in A-T.

Early PEG placement in A-T is beneficial in terms of safety and caregiver satisfaction. Late placement is associated with poor outcomes, thought to be due to factors including advanced lung disease and malnutrition.

REFERENCES

EVALUATION

For each question please choose which statements are true or false.

1. The following are all recognised causes of malnutrition in A-T
   a. Dysphagia and/or swallowing incoordination
   b. Insulin resistance and glucose intolerance
   c. Infections and an associated hyper-catabolic state
   d. Malabsorption
   e. Limited food intake with progressive disease

2. The following contribute to pulmonary morbidity in A-T
   a. Immune deficiency
   b. Pulmonary aspiration
   c. Malnutrition
   d. Chemotherapy
   e. Mucus hyperresponsiveness

3. Gastrostomy insertion in A-T
   a. Is associated with high levels of care giver satisfaction
   b. Should be considered early and proactively
   c. Normalises growth failure in all instances
   d. Should only be undertaken when growth failure and aspiration are present

4. Pulmonary aspiration in A-T
   a. Is present in over a quarter of case
   b. Can be silent aspiration
   c. More common in young children
   d. Cannot be assessed by videofluoroscopic examination