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Dealing with mealtimes and feeding in girls with Rett syndrome

Girls with Rett syndrome may commonly have difficult meal times. A number of factors contribute to this difficulty. The interplay between these factors may be complex and lead to a vicious circle the result of which may be inadequate growth, micro-nutrient deficiency, aspiration pneumonitis, oesophagitis, prolonged mealtimes or bloating. The evidence for the efficacy of available interventions is limited.

Pathological Processes

Dysphagia in all phases of swallow is frequent in Rett syndrome. This may result in slow meals with poor intake, with resulting poor growth or micronutrient deficiency. Poorly co-ordinated swallow may result in aspiration which may be symptomatic or clinically silent.

Poor growth is most frequently the result of inadequate nutrient intake. However some girls with Rett syndrome appear on history to have adequate intake and yet grow poorly.

Gastro-oesophageal reflux both symptomatic and asymptomatic may occur. This can result in aspiration of refluxed material or may lead to oesophagitis. Oesophagitis may result in discomfort at meals times which may inturn limit intake. It may also contribute to gut dysmotility, which may worsen the gastro-oesophageal reflux and contribute to discomfort from bloating.

Air swallowing in Rett syndrome is well documented and may result in bloating and discomfort or may potentially worsen gastro-oesophageal reflux by increasing intragastric pressure. Air swallowing is usually found in association with hyperventilation and apnoea.

Clinical Approach

A careful history and examination may give clues to the presence of each of these factors but this may need to be complemented by investigations or empirical trials of medications to sort out the relative importance of each.

Growth failure will be apparent from dietary history and measures of height and weight. This may be supplemented by more detailed anthropometric measures if required. Respiratory symptoms especially in association with a history of respiratory morbidity should prompt investigation for aspiration. Inadequate growth, mealtime discomfort, frequent vomiting or re-swallowing should prompt investigation for gastro-oesophageal reflux or where appropriate, an empirical trial of gastro-oesophageal reflux therapy. History of air swallowing with apparent discomfort and with or without the finding of resonant abdominal distension may suggest the possibility of a contribution of air swallowing to mealtime difficulties.

Dysphagia may respond to changes in feed posture or consistency or increase in calorie density of feeds. Gastrostomy with or without fundoplication may be required for either poor nutrition or recurrent aspiration. Micronutrient deficiency may be addressed by supplementation. Oesophagitis may be treated with ant-acid medication or fundoplication. Air swallowing has been treated with anxiolytics or topiramate. Establishing the efficacy of the various treatments used remains a significant challenge.