

The Significance Of Enamel Erosion As An Indication Of “Gastro-Oropharyngeal Reflux Disease” (GORD) In Adults With Intellectual And Developmental Disabilities (IDD)

H Hood, P May

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Abstract

Adults with intellectual and developmental disabilities (IDD) experience a high incidence of Enamel Erosion (EE), Gastroesophageal Reflux Disease (GERD), Oropharyngeal Dysfunction (OPD), and Chronic Lung Disease (CLD), all of which can contribute to increased morbidity and mortality. It appears that in patients with IDD, these conditions are often interrelated, which makes their evaluation and management more complex. Various health professionals, including gastroenterologists, otolaryngologists, pulmonologists, dentists, and speech therapists may be involved, often in a non-collaborative and uncoordinated manner which fails to take into account how one of these conditions may influence another. In adults with IDD, the presence of GERD, OPD, CLD and EE may be “syndromically” related which would suggest a need for improved collaboration between specialists traditionally focused on only one of these health conditions. We propose to designate the co-occurrence of EE, GERD, OPD, and CLD in an individual with IDD as “Gastro-Oropharyngeal Reflux Disease” or GORD. Furthermore, because of the ease of diagnosis of enamel erosion, the dentist is in the position to be most likely to determine an early diagnosis which might lead to prevention of life-threatening medical complications.

INTRODUCTION

Persons with intellectual and developmental disabilities (IDD) are living longer now, and most currently receive health services from generic, community-based health systems. Nonetheless much remains to be learned regarding the optimal evaluation and management of those health conditions that frequently occur in this medically complex patient population, a group that was also designated as a medically underserved population (MUP) by the American Medical Association in 2010 (Resolution 805-I-11).

The few studies that exist suggest that gastroesophageal reflux disease (GERD) is probably one of multiple, frequently-occurring health conditions experienced by persons with intellectual and developmental disabilities (1). Overall, in patients with IDD, the prevalence of esophagitis secondary to gastroesophageal reflux appears to be about 50 % (1).

It should be pointed out that esophageal manifestations (2) of gastroesophageal reflux, i.e. esophagitis, Barrett's

esophagus, and stricture are not the only clinical manifestations of gastroesophageal reflux disease. In addition to the esophageal manifestations of GERD, there are also so-called “extra esophageal” (or atypical) manifestations of GERD that extend beyond the esophagus (3-7). This is true in the general population as well as individuals with IDD. Examples of atypical manifestations of GERD encountered specifically in persons with IDD include enamel erosion (1,8), pulmonary manifestations such as laryngitis, reactive airway disease/asthma, and pulmonary fibrosis (9), and abnormal maladaptive behavior (10).

In general, proton pump inhibitors (PPI's) are very effective treatment of esophageal manifestations of GERD, but similar efficacy of PPI's for treatment of the extra-esophageal manifestations of GERD are not as evident. This appears to be true in persons with ID as well as the general population (10,1). Treatment of the extra- esophageal manifestations of GERD (enamel erosion, laryngo-pulmonary and behavioral) remain problematic in large part due to the fact that these

extra-esophageal manifestations are not specific for GERD, and may be due to other conditions as well.

It appears that if acidic refluxate extends beyond the superior esophageal sphincter into the mouth or hypopharynx, then both increased enamel erosion, hypopharyngeal inflammation and microaspiration/pulmonary dysfunction are more likely to occur (especially when associated with oropharyngeal dysfunction). Enamel erosion can be easily diagnosed by routine dental examination and hypopharyngeal inflammation can be readily determined by laryngoscopy by an ENT Specialist or fiber-optic endoscopic evaluation of swallow (FEES) by a Speech Therapist/SLP.

Chronic/acute lung disease is probably the most serious extra-esophageal manifestation of GERD and is a leading cause of death in persons with medically complex intellectual disabilities (MCID). Swallowing studies in children with feeding difficulties, also a clinical problem frequently observed in adults with intellectual disabilities, have shown a high frequency of pulmonary aspiration. Weir, et al (11), demonstrated in a series of 300 children with swallowing disorders that 34% demonstrated pulmonary aspiration on videofluoroscopic swallow study, which was silent in 81%. Silent aspiration was significantly associated with neurologic impairment and developmental delay. In addition, a relationship between asthma and enamel erosion has also been demonstrated (12).

Regarding the problem of pulmonary aspiration of refluxed material, it is important to determine the origin of the refluxate. Refluxate from the stomach which appears in the lungs will be acidic (so-called laryngopharyngeal reflux or LPR), but refluxate of swallowed food or liquid from the upper esophagus due to esophageal dysfunction will not be acidic (so-called esophagopharyngeal reflux). Thus treatment of pulmonary symptoms (e.g. cough, wheezing, etc) with proton pump inhibitors (PPIs) may work for the former (gastric), but not for the latter (esophageal). Also, pulmonary aspiration of saliva which is normally alkaline, may cause pulmonary symptoms (cough, wheeze), but would not be improved with PPI treatment. On the other hand, PPI treatment may benefit pulmonary symptoms caused by aspiration of refluxate from rumination (common in severe ID) or bulimia.

Lee, et al. (13), reviewed the problem of “silent” (no pulmonary symptoms) micro-aspiration of oropharyngeal or

gastric contents and concluded there was a strong association between gastroesophageal reflux and idiopathic pulmonary fibrosis. Others have noted that proximal esophageal non-acid reflux is common in patients with idiopathic pulmonary fibrosis, but that acid suppression therapy with PPI's, not surprisingly, had no effect on the symptom of cough. Thus it appears that chronic aspiration of any foreign substance may ultimately lead to lung damage, but clearly stomach acid is most damaging and more likely to lead to overt symptoms and chronic lung disease.

THE IMPORTANCE OF ENAMEL EROSION

Enamel erosion appears to be an important diagnostic sign which predicts not only the presence of GERD, but also an increased risk for pulmonary micro-aspiration of acid and/or other gastric substances known to be toxic to pulmonary tissue. Thus, in addition to being a sign or “risk factor” for GERD, enamel erosion may also be a “risk factor” for chronic pulmonary disease/dysfunction. This perspective is supported by the findings of Wang et al. 2010 (14), who demonstrated that in GERD patients, enamel erosions are more common in those patients with pulmonary symptoms than in GERD patients who do not have pulmonary symptoms. In other words, GERD patients who do not have enamel erosions are less likely to demonstrate pulmonary symptoms than GERD patients who do have enamel erosion. It seems apparent that pulmonary aspiration of either acidic or non-acidic substances can lead to pulmonary symptoms. While acid suppression therapy may not be able to suppress reflux or aspiration, it could be argued that acid-free reflux is less toxic to the lungs and therefore Proton pump inhibitors (PPI's) should be used to minimize the destruction that might occur from ongoing chronic aspiration of more acidic refluxate. Thus if enamel erosion is observed in the setting of chronic pulmonary symptoms, it might be reasonable to continue to treat with PPI's, even if cough continues in spite of acid suppression.

In a double blinded, randomized, placebo-controlled study, Wilder-Smith et al. (15) demonstrated efficacy of esomeprazole (Nexium) on the treatment of enamel erosions in a series of patients with GERD-related enamel erosion. The authors measured optical reflectivity (OR) using optical coherence tomography (OCT) before and after a three-week course of 40 mg per day esomeprazole (a proton pump inhibitor) in 30 patients versus controls. Optical reflectivity is a measure of enamel demineralization; the higher the OR the worse the demineralization/erosion. The esomeprazole

group (compared with the placebo control group) demonstrated significantly lower OR, which indicated that acid blockade may be useful in counteracting progression of GERD-related enamel erosion and lowering risk for reflux - related pulmonary damage in some patients.

If PPI treatment of enamel erosion were shown to be effective, this would indicate less Oral Acidity (OA) and a reduced risk of pulmonary damage should oral contents be aspirated.

THE IMPORTANCE OF OROPHARYNGEAL DYSFUNCTION

Micro-aspiration into the lungs would be even more likely to occur if any type of reflux were associated with oropharyngeal dysfunction (dysphagia) or any difficulty swallowing (11). Thus refluxate from the stomach could appear in the mouth first and then move from the mouth to the airway, or could flow from the upper esophagus directly into the laryngopharynx and lungs. Fiber-optic Endoscopic Evaluation of Swallowing Study (FEES) could provide evidence for this possibility even in the absence of enamel erosion

CAUSES OF ENAMEL EROSION OTHER THAN ACID REFLUX

Finally, it should be emphasized that demineralization can also be caused by a variety of acidic foods and liquid substances as well as environmental exposures. In addition, other factors to consider are the salivary pH and flow rate (i.e. the buffering capacity of saliva). In persons with medically complex intellectual and developmental disabilities anti-cholinergic psychotropic medication is frequently used and this can reduce salivary flow leading to a dry mouth and more acidic saliva. Thus patients with medically complex intellectual and developmental disabilities (MCID) may have multiple conditions that contribute to high oral acidity and an increased risk for enamel erosion and lung damage from reflux aspiration that should be considered in addition to GERD related enamel erosion.

SUMMARY AND CONCLUSIONS

Dangerous levels of oral acidity (OA) coupled with oropharyngeal dysfunction (OPD) increases the risk for micro-aspiration of acidic oral fluid into the airways which may increase risk for pulmonary fibrosis. This may be a common scenario in aging adults with medically complex intellectual disabilities. Evaluation of the salivary pH for

level of acidity, examination of the teeth for enamel erosion, and fiber-optic endoscopic evaluation of swallowing study (FEES) may be good objective screening procedures for the existence of a dangerous OA-OPD combination. If oral acidity and oropharyngeal dysfunction appear to be present, their etiologies need to be determined. First, the excessive use of carbonated beverages and/or acidic fruit juices needs to be ruled out. Secondly, drugs that affect salivary gland and oropharyngeal function need to be reconsidered. Neuroleptic (antipsychotic) and other drugs are well known to cause oropharyngeal dysfunction as well as impairment of normal alkaline saliva production which causes dry mouth and increased oral acidity. These drugs are frequently used in adults with medically complex intellectual disabilities to control behavior. Alternative drugs and/or behavioral programs can often be as effective and should probably be tried in the setting of combined oral acidity and oropharyngeal dysfunction.

At the very least a psychiatrist experienced in managing behavior problems in adults with medically complex intellectual and developmental disabilities should be consulted regarding the risk-benefit ratio of the continued use of neuroleptic drugs in an individual with medically complex IDD who has enamel erosion from oral acidity and evidence of oropharyngeal dysfunction and chronic lung disease. In addition to the psychiatrist, a pulmonologist will be needed to evaluate pulmonary function, a gastroenterologist to evaluate for gastroesophageal reflux disease (GERD), and an otolaryngologist to evaluate for laryngopharyngeal reflux (LPR).

In conclusion, enamel erosion is a common finding in adults with medically complex intellectual disabilities which significantly indicates: 1) a dangerous level of oral acidity (OA), 2) increased likelihood of gastroesophageal reflux disease and, 3) increased risk for pulmonary fibrosis.

When OA is considered in combination with oropharyngeal dysfunction (which is also common in adults with medically complex intellectual disabilities) pulmonary micro-aspiration of acidic fluid is more likely to occur, and, with time, may subsequently lead to pulmonary fibrosis and chronic pulmonary dysfunction. In order to effectively evaluate and manage these problems, collaboration of numerous health professionals who do not normally interact is needed. However if health systems can be developed to comprehensively address these problems, better health outcomes and an improved quality of life will be more likely

to occur for persons with medically complex intellectual and developmental disabilities.

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Author Information

Henry Hood, DMD

Lee Specialty Clinic; University of Louisville: School of Dentistry
Louisville, KY USA

Philip May, MD

Lee Specialty Clinic; University of Louisville: School of Medicine
Louisville, KY USA