SACCHARIN TEST OF MUCOCILIARY CLEARANCE IN KARTAGENER'S SYNDROME

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ABSTRACT

Saccharin test is a simple method for estimating mucociliary clearance in the upper respiratory passage. Its application in a clinically diagnosed patient with Kartagener's syndrome is reported.

Introduction

Primary ciliary dyskinesia, also known as Immotile cilia syndrome is now known to be a syndrome with many manifestations with the basic defect of d&ordered ciliary motility. Kartaaener's syndrome comprises absent or inflamed frontal sinuses, situs inversus with dextrocardia and bronchiectasis and is now considered a manifestation of immotile cilia syndrome. A simple bedside method of diagnosing reduced mucociliary clearance in these disorders has been reported. We report its application in a patient clinically and radiologically diaqnosed as Kartagener's syndrome.

Case Report

A 45 year old housewife presented to the chest clinic with nasal obstruction, running nose and cough with expectoration off and on for nearly 30 years. She had experienced intermittent fever for the past year. She had undergone a right lower lobectomy in 1970 for "recurrent couqh with copious expectoration". Full details were not available. She has had no past history of measles or whooping cough and denied any food or foreign body aspiration. On examination she was found to be pale, thin and without digital clubbing. Her trachea was shifted to right side and she exhibited fine to medium bibasilar crepitations which persisted after coughing. Heart sounds were better heard on the right chest and the liver dullness was absent on the right side. The central nervous system appeared clinically normal. On nasal examination, septal deflectjon to right with bilateral hypertrophied turbinates were seen. There was mucopus on the nasal floor on the right side. The cords appeared normal on indirect laryngoscopy. Roth tympanic membranes were retracted. Urine analysis revealed no casts, no albumin and no sugar. The peripheral blood count and erythrocyte sedimentation was normal. She exhibited a moderate iron deficiency anaernia. Acid fast bacilli were not seen on sputum smear (concentration method) on three days. Mantoux (with PPD) revealed 0 mm induration at 48 hours.

Skiagram paranasal sinuses revealed bilateral maxillary haziness (with mucosa of five mm +) and absence of frontal sinuses. The skiagram chest (PA view) revealed dextrocardia, tramlining and linear or irregular opacities left lower zone and fundus gas beneath right dome (Fig I). Bronchogram revealed predominantly tubular bronchiectasis of the left lower lobe.

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Fig 1. Skiagram chest PA view showing evidence of dextro cardia, fundus shadow on the right side, and bronchiectatic changes left lower zone.

Saccharin Test

A small grain of saccharin (approximately 1/10th of a commercially available tablet visually measured) is placed (with the help of a pair of forceps) two centimetres from the free edge of ala nasi on the inferior turbinate. The patient sits up during the entire period of testing. The patient is asked to indicate to the investigator, as soon as he/she appreciates the sweet taste of saccharin. Most normals report the taste within thirty minutes. The patient under report took one hour and fifty two minutes.

Discussion

Aufzelius reported abnormal respiratory mucosal ciliary activity in association with Kartagener's syndrome. He and his team have reported extensively on abnormal cilia in several syndromes [1, 2, 3, 4]

Mucociliary clearance is impaired in any disorder affecting the ciliary structure and function and this leads to recurrent or chronic respiratory infections. The schematic representation of the electron microscopic appearance of a cross section of a cilium is shown in Fig.2. Most reports pertain to abnormality or absence of the dynein arms. Functional ciliary dyskinesia has also been reported: Other reported abnormalities include absence of connecting spokes, deletion of control tubules, missing tubules, extra-tubules, compound cilia and intracytoplasmic cilia. These are all considered to be due to mutation.

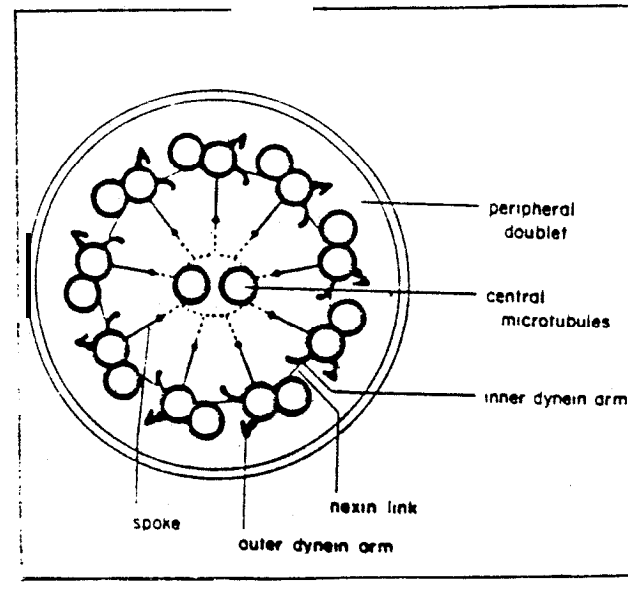

Fig 2. Diagram of ultra structure of respiratory cilium.

Even when the dynein arms are absent, these cilia exhibit some movement - they are not totally immotile. This has led to the use of the term - primary ciliary dyskinesia, which is gaining wide acceptance.

Ciliary motility has been, measured by photometry [5]*. Stanley et al studied 16 patients with primary ciliary dyskinesia and found that all had saccharin time of more than 60 minutes 14 of the 16 patients showed sinus mucosa of more than 5 mm thickness.*

The prevalence of primary ciliary dyskinesia is estimated to be between 1 in 15,000

to 1 in 30,000 live births (Rott[6])

Saccharin test for mucociliary clearance of the upper respiratory tract may be used as a screening test for primary ciliary dyskinesia syndrome of which Kartagener's syndrome is a subset. Electron microscopy for ultrastructural defects remains a research tool and is not available in most parts of the country.

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