Impact of Otitis Media in Pediatric Primary Ciliary Dyskinesia

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Conclusions
Primary ciliary dyskinesia (PCD) patients are greatly afflicted by otitis media (OM), and the majority receive multiple sets of pressure-equalizing (PE) tubes. Given the preponderance of otorrhea and clogged tubes, and frequent need for medical intervention but rarity of other otologic complications, PE tubes should be used judiciously. No clear correlations exist between genotypes and severity of disease in our cohort.

Introduction
Primary ciliary dyskinesia (PCD) is a ciliopathic, autosomal recessive genetic disorder that causes defective mucociliary clearance throughout the respiratory tract. We wish to explore the impact of otitis media (OM) in this population.

Project Goals
1. Evaluate the frequency of OM
2. Evaluate the frequency of OM complications and surgical interventions.
3. Establish a treatment paradigm for this cohort.

Methods
A retrospective chart review of patient registry at the PCD Center of an academic tertiary children’s hospital. Demographic, clinical and audiologic information were analyzed.

Patients with a known diagnosis of PCD were selected from our database. Diagnosis was confirmed on either (1) presence of a known causal genotype (2) presence of characteristic structural defects on electron microscopy (3) a compelling clinical picture concomitant with cystic fibrosis exclusion.

Results

Patient Characteristics
- N = 54 (35.2% Male, 64.8% Female
- Age at diagnosis
  - Mean: 5.24 years
  - Median 4 years
  - Range: 0-18 years
- Ethnicity

Audiometric data
- Available for 19 Patients
- Hearing loss was present in 32 ears (84%)
  - 24 were conductive in nature
- Mean speech reception thresholds:
  - 23.2dB on the right
  - 22.4dB on the left

Results Continued

Treatment
(A) Tube placement
- Otitis media with effusion was diagnosed in 51 patients (94%) and 36 (70.6%) received tubes
- 27 (75%) received multiple PE tubes.
- 26 (72.2%) had otorrhea or clogged tubes

(B) Other procedures
- Four patients needed tympanoplasty and two received mastoidectomy.

(C) Admissions
- Four patients required 6 ear-related admissions for intravenous antibiotics for the following indications:
  - Following placement of PICC and tubes (chronic otorrhea) 1
  - From clinic (chronic otorrhea) 1
  - Post-op observation after tube removal (chronic otorrhea) 1
  - Otorrhea and low PFTs 1
  - Resistant OM and sinusitis 1
  - Resistant OM 1

Genetics
Genetic data was assessed in light of otologic disease:

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<th>Genotype</th>
<th>% Yes</th>
<th>% No</th>
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*HL = hearing loss, AOM = acute otitis media, OME = otitis media externa