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Title: PCD – As serious as CF in every day lung clinic?

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Body: Background: Cystic fibrosis, CF, and Primary cilia dyskinesia, PCD, have very different genetic background, but present very similar in clinic with vicious mucus, bacteria, bronchiectasis and negatively affected lung function. Our regimen involves mucolytica, inhalations, airway clearance and anti-bacterial treatment regardless of diagnosis, but in all cases individualized. Aim of study: to see whether PCD patients as a group were as affected as CF patients. Patients: All patients, CF and PCD, seen regularly at our clinic were compared as a whole group and also in an age and gender matched subgroup of 21 pairs. Results: When comparing lung function in the two groups, $FEV_{1,}$ (in percentage of expected for age and length) showed, to our surprise, to be worse for the PCD group, both in the main group and in the subgroup. The bronchiectases are evaluated separately and will be presented at the Conference. There were more, but not exclusively, pseudomonas infections in the CF group – 38 vs 14 %. There was no B.cepacia or S.maltophilia in the PCD group. The BMI was as expected a little lower in the CF group. Conclusions: The more affected lung function amongst the PCD patients could be due to later diagnosis and less importance given to lung treatment. The focus on early diagnosis, effective inhalation and treatment regimen in the CF group have resulted in better results despite the very complex and severe disease.