

## Reviewer's report

**Title:** Primary Ciliary Dyskinesia (Siewert's / Kartagener's Syndrome): Respiratory symptoms and psycho-social impact

**Version:** 1 **Date:** 29 September 2003

**Reviewer:** Liesl L Osman

### Reviewer's report:

#### General

This is an interesting paper describing quality of life impact of ciliary motility disease. The paper is well written, and data is competently analysed and presented. I have two concerns/criticisms. First, there is an extent of overkill through the presentation of too much data. This is a pity, because the contention of the authors that early diagnosis of PCD has marked benefit is an interesting message. Second, the paper needs to discuss and justify why and how early diagnosis of PCD can have so much benefit - I think it is quite striking that so much benefit should (apparently) be gained by early diagnosis of an illness for which there is really no very intensive treatment, and where the respondents are presumably drawn from a range of clinics with different approaches.

I would like some data on ongoing treatment of the people in the survey. - eg how many antibiotic courses they have had in the past year. I see that Bisgaard's clinical treatment is quite aggressive with regard to antibiotics - what evidence do they have that this is the case in the UK? Ditto physiotherapy. I would be sceptical that people who really have quite good lung function would be practising physiotherapy every day among patients attending UK clinics.

If they don't have any data I think they should at least acknowledge this and make some comment or just how PCD is managed in UK clinics. What they mean by prophylaxis should be spelt out clearly.

My concern is, that there might be a statistical artefact explaining the apparent benefit of early diagnosis, a cohort effect in that people who had early diagnosis are all young and people who had late diagnosis are significantly older. I understand that they have adjusted for age, but feel that there may not be sufficient variation within the two groups to make this adjustment adequate. I would like some comment and justification that this is not the case.

#### Discretionary Revisions (which the author can choose to ignore)

1. I think Figure 1, and figure 6 are unnecessary, as are sufficiently reported in text.
2. P7 could remove family history para - irrelevant to discussion.
3. P10 Remove table 2 and provide a shorter para simply stating that there were no differences between PCD -SS and PCD \_ SI, as shown in figs 2,3,4
4. Figure 7 is unnecessary, it adds nothing to the simple statement in para 1 of the discussion.
5. Paper would benefit by statement on incidence of PCD in the UK

Minor Compulsory Revisions (such as missing labels on figures, or the wrong use of a term, which the author can be trusted to correct)

Major Compulsory Revisions (that the author must respond to before a decision on publication can be reached)

1. As above, needs comment on clinical management of PCD in the UK, and more discussion and justification of benefit of early diagnosis for patients.

**Advice on publication:** Unable to decide on acceptance or rejection until the authors have responded to the major compulsory revisions

**Level of interest:** A paper whose findings are important to those with closely related research interests

**Quality of written English:** Acceptable

**Declaration of competing interests:**

None