Reviewer’s report

Title: Primary Ciliary Dyskinesia in Japan: Systematic review and meta-analysis

Version: 0 Date: 15 May 2019

Reviewer: Guillaume Thouvenin

Reviewer’s report:

Importance and originality

Is the question posed original, important and well defined? Yes

What does the work add to what is already in the published literature? It add data about a specific population, not well studied

Scientific reliability

Is the study design appropriate to the question? Yes

Are the participants adequately described, defined and representative of the intended population? Yes

Are inclusion and exclusion criteria described? Yes

Are the methods appropriate and well described? Yes

What are the strengths and weaknesses of the methods?

Is the main outcome measure clear? Observational study : no main outcome

Does the study follow the appropriate EQUATOR Network reporting guidelines?

- Randomised trials - CONSORT
- Observational studies - STROBE
- Systematic reviews - PRISMA Yes
- Case reports - CARE
- Study Protocols - SPIRIT
- Animal pre-clinical studies - ARRIVE
Are the data sound and well controlled?

Is the interpretation well balanced and supported by the data?

Are the title and abstract an accurate description of the work? Yes

Are the references up to date and relevant? Yes

Can the writing, organization, tables and figures be improved?

Tables not at the end of the text?

Table 1 and text above: Classification of the items in sub group (by example, infertility between fever and hemoptysies / mix of ENT and respiratory items in medical history)

Not sure about the interest of comparison of >18 and <18 yrs.

Absence of chronic bronchitis? Absence of rhinitis?

9.31E-07?

Text not justified line 176 - 185

Age of diagnosis not defined (criteria?).

Table 2 (and text above) not well presented.

not very understandable

FEV₁

Values unit are missing (Age, nasal NO, ...)

Data mixed, with different statistical analysis...

Writing IDA defects only is not appropriate (IDA defect with microtubule disorganization is better)

Do not mention

Table 3 and text above not well presented.

Do not mix results and biopsy sites

IDA defects alone? Not clear
"IDA defect may include IDA defect with microtubule disorganization 232 (MTD), not widely recognized until causative genes were identified in 2013 [18]."

Discussion not results

Table 4

I am not able to analyse the statistical analysis...

If it is not correct, I would prefer to read comparison with international litterature data in discussion part.

No nNO results ?

Genetic testings in other diagnostic tests ?

"11 patients died of respiratory infection" : particular severe patients ? (not usual in PCD patients)

Discussion

Too long sentences

Interesting ideas but confusing discussion.

Difficult to understand the milder symptoms hypothesis and the rate of death, particularly in the pediatric population.

Are there any ethical or competing interest issues you would like to raise?

Are the methods appropriate and well described?
If not, please specify what is required in your comments to the authors.

Yes

Does the work include the necessary controls?
If not, please specify which controls are required in your comments to the authors.

Unable to assess

Are the conclusions drawn adequately supported by the data shown?
If not, please explain in your comments to the authors.

Yes
Are you able to assess any statistics in the manuscript or would you recommend an additional statistical review?
If an additional statistical review is recommended, please specify what aspects require further assessment in your comments to the editors.

I recommend additional statistical review

Quality of written English
Please indicate the quality of language in the manuscript:

Not suitable for publication unless extensively edited

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