Reviewer’s report

Title: First clinical experience with the Kora pacemaker system in congenital complete heart block in newborn infants

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Reviewer: Manfred Marx

Reviewer's report:

The article is well-organized, the sections are well-developed, providing a short introduction, the methodology is clearly explained. Results are well-written and easy to understand, tables or figures aid understanding and provide more detailed information.

The main question addressed by the authors - adequate growth and development - is highly relevant and interesting not only for the pediatric cardiology expert.

Page 2

Conclusion

The use of a pacemaker system with a maximum upper rate interval of 95 beats per minute in infants suffering from congenital complete heart block and showing high aortic VTI values seem to be sufficient for adequate growth and development.

The question is: what is adequate??? All neonates grew along their percentile curve: although it is difficult so extract exact numbers from the percentile curves, all infant grew along the 10th percentile (either in height or in weight): per definition: this is underweight.

Page 3

Authors hypothesize that the Kora pacemaker system allows for adequate growth and development in newborns and infants suffering from CCHB, as cardiac output adaption took place prenatally.

The neonatal high aortic velocity time integral (VTI) might be an adaption mechanism parameter, however it would be highly interesting to see, how VTI develops during follow up.
The appropriateness of MV sensors in children with pacemakers has been shown by Cabrera et al:

Thirty-eight healthy children, ages 6-14, underwent a treadmill maximal exercise test. Subjects were divided based on body surface area (BSA) and MV rate response parameters were selected. Respiratory rates and tidal volumes were entered into the Kappa 400 rate response algorithm to calculate sensor-driven rates.

This paper reported the results of a computer simulation using healthy children, and it is more than a question whether this mode works with epicardial leads???

Assuming that the minute ventilation sensor function would work with epicardial leads, the maximum stimulation frequency cannot be more than 95/min - which might be definitely too low, if spontaneous heart frequency would never be higher than this.

Looking at the three circadian heart rate diagrams recorded at 2.5, 2 and 1.9 years (!) respectively, all 3 patients do have an spontaneous escape rhythm which is higher than the stimulation rate - at least during daytime. There is no information about the stimulated frequencies during the years before. The question whether the limited stimulation frequency alone is responsible for reduced growth can only be answered with a control group of infants paced with higher frequencies.

In conclusion, the major limitations of this study are

The definition of adequate growth must be defined precisely. If we do not accept the reported results as good enough then industry would be forced to provide devices for small infants with adequate stimulation frequencies.

There is no evidence that the minute ventilation sensor function also works with epicardial leads.

There is no evidence in the paper that the minute ventilation sensor function was helpful

There is no matched control group with infants paced with higher frequencies.

With a change of perspective this paper should be accepted for publication to force industry to provide better devices for these small patients - they do have the right for that!

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.

No

Are the conclusions drawn adequately supported by the data shown?
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No

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Not relevant to this manuscript

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