Reply to the comments by Dr. Udayakumaran on our article “A late complication of CSF shunting: acquired Chiari I malformation”

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Dear Editor:

We are grateful to Dr. Udayakumaran for his interest in our paper [1] on secondary Chiari I malformation developing in patients shunted for congenital hydrocephalus or cyst.

As described in the article, we interpreted this anomaly (“acquired” because in no case it was present at the time of the initial diagnosis and treatment) as the result of CSF venting from the intracranial, mainly supratentorial, space with secondary reduction of the intracranial volume and consequent “bidirectional” herniation of the cerebellum, downward into the cervical canal and upward into the tentorial notch. CSF shunt function was described as “satisfactory” with reference to the correction of the ventricular dilation, obviously not with reference to the tonsil herniation.

In our opinion, disproportion between the exceeding cranial volume and the reduced intracranial content is sufficient to explain the novel appearance of this condition, without the necessity to take into consideration the slit ventricle syndrome. Everyone experiences cases of slit-like ventricles, with or without clinical manifestation; anyway, tonsillar herniation is quite exceptional in these cases.

Although the adopted surgical strategy was, for some regards, identical to that traditionally adopted for the treatment of the slit ventricle syndrome, this does not imply that the anomaly described is comparable to that.

Coming to the case proposed for discussion and comparative analysis, we have some difficulties in considering that tonsillar herniation observed by the author similar to our own cases. In his case, the radiological picture suggests a poorly functioning CSF shunt with secondary tonsillar herniation, rather than a satisfactorily working shunt (as described in our cases).

Certainly this topic remains matter of debate and further contributions are appreciated to highlight this intriguing condition in shunted chronic hydrocephalic subjects

References

1. Caldarelli M, Novegno F, Di Rocco C (2009) A late complication of CSF shunting: acquired Chiari I malformation. *Childs Nerv Syst* 25(4):443–452

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