Incidence and type of bicuspid aortic valve in two model species

Fernández M.C.\textsuperscript{1,2}, López-García A.\textsuperscript{1,2}, Soto M.T.\textsuperscript{1}, Durán A.C.\textsuperscript{1,2}, Fernández B.\textsuperscript{1,2}

\textsuperscript{1}Department of Animal Biology, Faculty of Science, University of Málaga, Spain
\textsuperscript{2}Biomedical Research Institute of Málaga (IBIMA), University of Málaga, Spain

Bicuspid aortic valve (BAV) is the most frequent human congenital cardiac malformation, with an incidence of 1-2\% worldwide. Two morphological types exist: type A (incidence $\sim$0.75-1.25\%) and type B (incidence $\sim$0.25-0.5\%), each with a distinct aetiology and natural history. Currently, ten animal models of BAV have been described in two different rodent species: one spontaneous Syrian hamster (\textit{Mesocricetus auratus}) model of BAV type A and nine mutant laboratory mouse (\textit{Mus musculus}) models of BAV type B. It remains to be elucidated whether the mutations leading to BAV in these models are type-specific or whether there are inter-specific differences regarding the type of BAV that hamsters, mice and humans may develop.

To solve this issue, we have characterized the incidence and types of BAVs in four inbred, two outbred and two hybrid lines of Syrian hamsters (n=4,340) and in three inbred, three outbred and one hybrid lines of laboratory mice (n=1,661) by means of stereomicroscopy and scanning electron microscopy. In addition, we have reviewed and calculated the incidence and type of BAVs in the published papers dealing with this anomaly in mice.

Our results indicate that the Syrian hamster develops BAVs type A and B including a variety of morphologies comparable to those of humans, whereas the mouse develops only BAVs type B with a short spectrum of valve morphologies. Thus, inter-specific differences between human and mouse aortic valves must be taken into consideration when studying valve disease in murine models.

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