Application of the US National Institute of Health (NIH) 2008 guidelines for von Willebrand Disease in a National Paediatric Comprehensive Care Centre.

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Introduction
Von Willebrand Disease (VWD) is considered the most common inherited bleeding disorder.
To improve clinical diagnosis of VWD, practise guidelines were published by the US National Institute of Health (NIH) in 2008. Patients with plasma von Willebrand factor (VWF) i.e. VWF:Ag, VWF:RCo or VWF:CB levels ≤0.3 IU/ml were diagnosed with ‘VWD’. Individuals with plasma VWF levels in the range 0.3IU/ml to the normal range were diagnosed with ‘Low VWF levels’.

Methods
In the current study, we retrospectively extracted the case records of children under 18 years on the national database with VWD or possible VWD. These records were then analysed according to the NIH diagnostic criteria.

Results
220 children had been historically diagnosed with VWD or possible VWD. Following the review there was a 68% reduction in the number of patients diagnosed with Type 1 VWD. Predictably, no change in numbers diagnosed with Type 2 and Type 3 VWD. 85 (38% of total population) is now classified as Low VWF. 35 (16% of total population) were deemed to have no form of VWD and reclassified as normal.

Conclusions
Our data strongly suggest over-diagnosis of VWD in this paediatric population. This review has eliminated possible VWD and unspecified VWD type categories. This has however implications for service resources where newly reclassified patients need to be informed and counselled. Also, there is a requirement to establish management strategies for patients newly diagnosed as low VWF at times of bleeding or at risk for bleeding.