PEDIATRICS

How should we classify DSD?

Two papers published in the *Journal of Pediatric Urology* have addressed the controversy surrounding the terminology used to describe patients with abnormalities of sex determination.

The nomenclature for such disorders has long been contentious. Patients and their families often struggle to come to terms with not just the diagnosis but also the difficulties associated with being dubbed 'intersex'. This term has been used since the 1920s, when it was adopted in favor of the historical labels 'hermaphrodite' and 'pseudohermaphrodite' which were considered derogatory and stigmatizing.

In 2005, a consensus group sought to resolve the issue by proposing a new terminology. They suggested that the umbrella term 'intersex' be replaced with 'disorders of sexual development', and that further classification be based on karyotype; that is, 46 XX DSD, 46 XY DSD, ovotesticular DSD, or 46 XX testicular DSD.

A team at Southampton University Hospital, UK, used a questionnaire to assess the opinions of parents of children with and without DSD, and of health-care professionals, on the terminology proposed by the 2005 consensus group. Overall, 86% of those questioned preferred the term 'disorders of sexual development' compared to 'intersex'. This rose to 95% amongst parents of a child with a DSD. All groups questioned felt that the new term would make it easier for parents to understand the condition, and to explain it to the affected child and their relatives. The more-specific categories 46 XX DSD and 46 XY DSD were considered confusing by 40% of those questioned, and 85% felt that these terms did not aid comprehension. By contrast, this terminology was popular amongst health-care workers.

This discrepancy was also recognized by Ian and Alistair Aaronson from the University of South Carolina. They have proposed that the nomenclature would be improved by basing it on histological integrity of the gonads, which is more clinically relevant than the underlying karyotype.

"The karyotype, although providing important information, does not get one very far with regard to the definitive diagnosis, several conditions having quite variable chromosome patterns," says Ian Aaronson. "By contrast, gonadal histology provides an excellent conceptual basis for understanding sex differentiation in the human embryo, and by extension, the various errors that may occur on these pathways leading to the various DSDs." Aaronson and Aaronson suggest that DSD subtypes be classified as ovarian, ovotesticular, testicular or dysgenetic, depending on gonadal histology. They stress, however, that biopsy of the gonads should only be performed when a diagnosis cannot be reached by extrapolating from biochemical and clinical data. Such a classification system might prove helpful not only to patients, who would be better able to understand the physiology of their condition, but also to junior medics and students, who often struggle to comprehend the role of the gonad in sex differentiation.

It remains to be seen whether either of these DSD classification tools are here to stay. Either way, the controversy surrounding the nomenclature most certainly is.

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Original articles Davies, J. H. *et al*. Evaluation of terminology used to describe disorders of sexual development. *J. Pediatr. Urol.* doi:10.1016/j.jpurol.2010.07.004 | Aaronson, I. & Aaronson, A. How should we classify intersex disorders? *J. Pediatr. Urol.* doi:10.1016/j.jpurol.2010.04.008