Classification Puts Childhood Vasculitis in Order

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A new international classification for childhood vasculitis promises to improve existing criteria by bringing them into line with current clinical practice, reported Dr. Seza Ozen and fellow associates.

The new criteria, endorsed by the European League Against Rheumatism and the Paediatric Rheumatology European Society, were needed to update existing criteria and make them more specific.

It is the first internationally agreed-upon classification of the vasculitides observed in children, and was devised by gathering opinions from a wide range of pediatric rheumatologists and nephrologists from around the world, reported Dr. Ozen of Hacettepe University, Ankara, Turkey, and associates (Ann. Rheum. Dis. 2006;65:936-41).

The working group of experts updated the classification of Henoch-Schönlein purpura, Kawasaki disease, childhood polyarteritis nodosa, Wegener's granulomatosis, and Takayasu arteritis.

"It was agreed that vessel size would form the backbone of the criteria," as had been decided at the Chapel Hill Consensus Conference for adult vasculitides, the investigators noted.

In addition, small-vessel vasculitis was divided according to the presence or absence of granulomas, "as this feature has important distinguishing implications." A category of "other vasculitides" was added for those disorders in which an etiologic process was defined or which did not fit into existing categories.

For Henoch-Schönlein purpura, the primary change was that palpable purpura is now a mandatory criterion. Biopsy findings also were clarified, and since arthritis is common in the childhood type of this disorder, it is now included in the criteria.

The existing criteria for Kawasaki disease were retained. But since coronary artery disease is so important in this disorder, children who have typical echocardiographic changes can now be classified as having Kawasaki disease even if they do not fulfill four of the remaining criteria.

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In addition, the perineal desquamation frequently noted in Kawasaki disease has been included under "changes in extremities."

"Pediatric data during the last 10 years have shown that polyarteritis nodosa in children is hard to classify using the definitions applicable to adults," Dr. Ozen and associates said. "We decided that abnormal angiography or a characteristic biopsy are mandatory criteria for the diagnosis of the disease" in children.

In addition, the criterion of hepatitis B surface antigen was eliminated, since this is no longer a major feature in pediatrics, "thanks to improving vaccination" practices. And since cutaneous disease is common in childhood polyarteritis nodosa, it has been added to the new criteria.

For Wegener's granulomatosis, subglottic, tracheal, or endobronchial stenosis has been added as a new criterion, because these are frequent features of the childhood disease.

Changes in technology prompted a revision of the criterion concerning radiographic imaging so that CT results are now included. And antineutrophil cytoplasmic antibody testing has also been added.

For Takayasu arteritis, the angiographic criterion was updated to reflect changes in technology, and it is now a mandatory criterion.

Hypertension is common and often is the only presenting sign in pediatric patients, so it has been included as a criterion even though it is nonspecific.

Overall, the new classification should benefit pediatricians and rheumatologists in practice, noted Dr. Ozen and associates. "We believe this was an important task, as appropriate classification criteria for vasculitis in children have been missing for far too long.

"The next step will be to validate these criteria using patients and control groups," they said. ■