

Diagnostic criteria for Proteus syndrome (PS)

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Table 1 Revised PS diagnostic criteria.

To make a diagnosis of PS, one must have all the general criteria, and various specific criteria

<i>General Criteria</i>	<i>Specific Criteria</i>
<p>All the following: Mosaic distribution of lesions Sporadic occurrence Progressive course</p>	<p>Either: Category A or, Two from category B or, Three from category C</p>
<i>Specific criteria categories</i>	
<p>A. 1. Cerebriform connective tissue nevus^a</p> <p>B. 1. Linear epidermal nevus 2. Asymmetric, disproportionate overgrowth^b One or more: (a) Limbs (b) Hyperostosis of the skull (c) Hyperostosis of the external auditory canal (d) Megaspondylodysplasia (e) Viscera: Spleen/thymus 3. Specific tumors before 2nd decade One of the following: (a) Bilateral ovarian cystadenoma (b) Parotid monomorphic adenoma</p>	<p>C. 1. Dysregulated adipose tissue Either one: (a) Lipomas (b) Regional lipohypoplasia 2. Vascular malformations One or more: (a) Capillary malformation (b) Venous malformation (c) Lymphatic malformation 3. Lung cysts 4. Facial phenotype All: (a) Dolichocephaly (b) Long face (c) Down slanting palpebral fissures and/or minor ptosis (d) Low nasal bridge (e) Wide or anteverted nares (f) Open mouth at rest</p>

This table is adapted from prior publications.^{6,8,21}

^aCerebriform connective tissue nevi are skin lesions characterized by deep grooves and gyrations as seen on the surface of the brain. See text and Figure 1 for details.

^bAsymmetric, disproportionate overgrowth should be carefully distinguished from asymmetric, proportionate, or ballooning overgrowth. See text and Figure 1 for details.