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Vasculitis in children

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Primary systemic vasculitides of the young are relatively rare diseases, but can have a significant morbidity and mortality. The purpose of this review is to provide an overview of the paediatric vasculitides. Vasculitides that predominantly affect children will be considered in more detail than vasculitic diseases that although are seen in children affect adults more commonly, such as the ANCA associated vasculitides. New classification criteria for childhood vasculitis have recently been proposed and are currently undergoing validation. Epidemiological clues continue to implicate infectious triggers in Kawasaki Disease and Henoch Schönlein purpura. Several genetic polymorphisms have now been described in the vasculitides that may be relevant in terms of disease predisposition or development of disease complications. Treatment regimens continue to improve, with the use of different immunosuppressive medications and newer therapeutic approaches such as biologic agents. However new challenges are looming in regards to the role of inflammation in endothelial health and the long term cardiovascular morbidity for children with primary systemic vasculitis. International multicenter collaboration is of utmost importance in order for us to further advance our understanding and improve the treatment and outcome of systemic vasculitis in the young.

Introduction

Primary systemic vasculitis (PSV) of the young is characterised by the presence of inflammation in the walls of blood vessels, with resultant tissue ischaemia and necrosis [1]. Apart from relatively

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common vasculitides such as Henoch-Schőnlein purpura (HSP) and Kawasaki disease (KD), most of the primary vasculitic syndromes are rare in childhood, but with a significant attendant morbidity and mortality. Latest developments in the field of paediatric vasculitis include: a) the recent consensus conference for a new international classification of childhood vasculitides b) the identification of various genetic polymorphisms that may be relevant in terms of disease predisposition or development of disease complications, and c) novel therapeutic approaches including the increasing use of biologics in cases where current standardized first line treatment fails to induce or sustain remission.

In addition, with the increased survival of affected patients, new challenges particularly in regard to longer term cardiovascular morbidity in children who survive vasculitis are looming. The purpose of this review is to present an overview of the primary vasculitides that more often affect children, and to review recent areas of both scientific and clinical importance.

New international classification of childhood vasculitis

There has been for many years a need for an acceptable classification of childhood vasculitis. So far there has been much reliance on adult vasculitis classification systems and definitions that are suboptimal for paediatric vasculitides. A recent International Consensus Conference held in Vienna in June 2005 under the auspices of the Pediatric Rheumatology European Society (PReS) resulted in a new proposal for childhood vasculitis classification summarised in Table 1 [2]. These criteria are currently being validated and it is anticipated that this will be an important step in providing a "fit for purpose" classification system to be used in future epidemiological studies and clinical trials of paediatric vasculitis.

Henoch Schönlein Purpura

HSP is the most common form of systemic vasculitis in childhood and predominantly affects small vessels [3]. The presence of palpable purpura in the absence of thrombocytopenia is an essential classification criterion.

Table 1
Proposed classification of childhood vasculitis (adapted from Ozen et al. 2006 [2]).

I.	Predominantly large vessel vasculitis
	Takayasu arteritis
II.	Predominantly medium-sized vessel vasculitis
	Childhood systemic polyarteritis nodosa
	Cutaneous polyarteritis nodosa
	Kawasaki disease
III.	Predominantly small vessel vasculitis
	A. Granulomatous
	Wegener's granulomatosis
	Churg Strauss syndrome
	B. Non-granulomatous
	Microscopic polyangiitis
	Henoch-Schonlein purpura
	Isolated cutaneous leukocyclastic vasculitis
	Hypocomplementaemic urticarial vasculitis
IV.	Other vasculitides
	Behçet's disease
	Vasculitis secondary to infection (including hepatitis B-associated PAN), malignancy and drugs including hypersensitivity vasculitis
	Vasculitis associated with connective tissue disease
	Isolated vasculitis of the CNS
	Cogan's syndrome
	Unclassified