

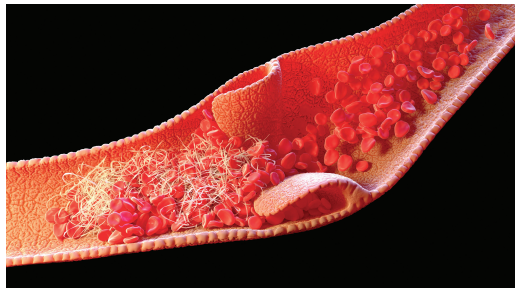
# Do you suspect SCPCD?\*

Don't miss  
the details

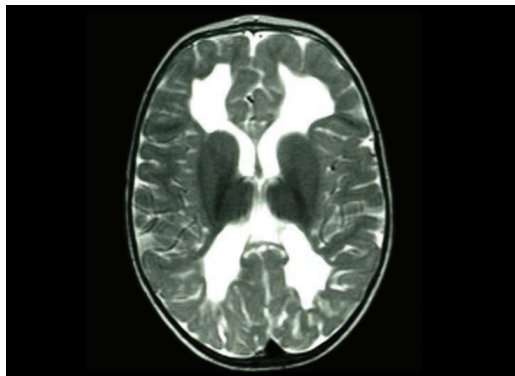
## SIGNS & SYMPTOMS

Symptoms are driven by a shift in the balance between pro- and anti-coagulation factors that result in excess coagulation<sup>3</sup>

Excess coagulation causes the following clinical features.\*



Getty images



Chalmers E et al. Arch Dis Child. 2011 Nov;96(11):1066-71.

### Venous thrombosis

Large vessel venous thromboses may also occur, e.g. renal vein thrombosis.<sup>2</sup>

### Cerebral venous thrombosis

Affected neonates also typically show significant neurological injuries resulting from antenatal or early postnatal cerebral venous thrombosis with secondary periventricular haemorrhagic infarction and hydrocephalus<sup>1</sup>. Neurological complications can occur *in utero* and be diagnosed antenatally.<sup>5</sup>

### Retinal vessel thrombosis

Blindness is a common manifestation of severe PC deficiency and may arise from vitreal bleeding, retinal vein, artery or vitreal vein thrombosis with retinal detachment manifesting as leucocoria or ischaemic optic atrophy.<sup>1</sup> Ophthalmic lesions may occur before or after birth.<sup>6</sup>



Chalmers E et al. Arch Dis Child. 2011 Nov;96(11):1066-71.

### Purpura fulminans

Purpura fulminans (PF) is a rapidly-progressing thrombotic disorder involving haemorrhagic infarction of the skin and disseminated intravascular coagulation (DIC).<sup>1</sup> At first, skin lesions appear dark red and subsequently become purple-black and indurated.<sup>1</sup> In SCPCD, these lesions develop at pressure points, such as the back of the head, buttocks and heels.<sup>3</sup>



BM 2018 BMJ Publishing Group Ltd & Royal College of Paediatrics and Child Health. All rights reserved. Reference: Sharma S. Archives of Disease in Childhood - F&N, 2015;100:F453. J

\*The symptoms presented here are not an exhaustive list. Other symptoms may be present in the event of severe PC deficiency.

If you suspect SCPCD conduct a  
PC Activity Assay Immediately<sup>1,3</sup>

\* SCPCD: Severe Congenital Protein C Deficiency

1. Chalmers E, et al. *Archives of Disease in Childhood*. 2011;96(11): 1066-1071.

2. Price VE, et al. *Semin Fetal Neonatal Med*. 2011;16(6):318-322.

3. Goldenberg N, Manco-Johnson M. *Haemophilia*. 2008;14(6): 1214-1221.

4. Tadmouri GO, et al. *Reproductive Health*. 2009;6(17):1-9

5. Kirkinen P, et al. *Prenat Diagn*. 2000;20(9):746-749

6. Ergenekon E, et al. *Br J Ophthalmol*. 2000;84:117-121.