

## PRACTICE



## PRACTICE POINTER

## Purpuric and petechial rashes in adults and children: initial assessment

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Bleeding into the skin or mucosa from small vessels produces a purpuric rash, or smaller petechiae (1-2 mm in diameter). Purpura is not a diagnosis but can be the presenting feature of serious conditions, such as meningococcal sepsis and acute leukaemia, which require urgent diagnosis and management. Equally, it can cause patients alarm but requires little more than a single assessment and reassurance. Differentiating between the two scenarios is important. This article focuses on recognition of the serious diagnoses and recommendations for urgent referral. Once such diagnoses have been excluded, other causes can be investigated or the patient managed by observation alone.

### Is the rash purpuric?

A cardinal sign of a purpuric rash is that it does not blanch on pressure, unlike exanthema, telangiectases, or allergic rashes. This sign of meningococcal sepsis has been the subject of public health campaigns to help parents recognise its importance and seek urgent medical attention (fig 1⇓).

It is crucial to assess for features of serious illness in all patients with purpura.

### What can cause a purpuric rash?

Patients with purpura can generally be divided into those who are acutely unwell and those who are not. Table 1⇓ outlines the causes of each. The rash may indicate reduced number or function of platelets, another bleeding diathesis such as von Willebrand disease, or defective supporting tissues. Thrombocytopenia is usually severe (platelets  $<20 \times 10^9/L$ ) before spontaneous petechial haemorrhages appear.

Purpura is an uncommon presenting problem because of the rarity of its more serious underlying causes. Very fine petechiae can accompany viral illnesses,<sup>1,8</sup> but the more serious causes still need to be excluded.

### Purpura in ill patients

In the most feared causes of a purpuric rash—meningococcal sepsis and acute leukaemia—the patient is usually unwell, often more acutely so with sepsis.

### Meningococcal sepsis

Although meningitis is the most common form of invasive meningococcal disease, meningococcal sepsis occurs without meningitis in 5-20% of invasive infections,<sup>2,3</sup> which have an overall incidence of about 1/100 000.<sup>9</sup> In meningococcaemia, mortality is up to 40%, even with appropriate antibiotic therapy. Most deaths occur within the first 24 hours.<sup>2,4</sup>

Most cases (40%) occur in pre-school children, especially infants, with further peaks in late adolescence and in people over 65 years.<sup>2,9</sup> Medical risk factors include asplenia, HIV infection, complement deficiency, and other immunosuppressed states. Purpura is secondary to disseminated intravascular coagulation. Bleeding may be secondary to depletion of platelets and coagulation factors from the consumptive coagulopathy. Other clinical features are listed in table 1⇓ and given in more detail in National Institute for Health and Care Excellence<sup>2</sup> and Scottish Intercollegiate Guidelines Network<sup>3</sup> guidelines.

### Acute leukaemia

The incidence of acute leukaemia is 6/100 000 in children, rising to 70/100 000 at 80 years.<sup>10</sup> Patients are not necessarily acutely unwell at presentation. Purpura or bleeding may have an acute or sub-acute onset and take the form of widespread petechial haemorrhages (fig 2⇓) or ecchymoses on the limbs and trunk. Table 1⇓ outlines the other clinical features.

**What you need to know**

- Assess all patients with suspected purpura for features of serious illness
- If invasive meningococcal disease is suspected, administer parenteral antibiotics immediately, but do not delay hospital admission
- In all other patients, severe thrombocytopenia must be excluded. Immediately refer children and young people for assessment; adults should have a full blood count and coagulation screen within 48 hours

**Purpura in patients who are not acutely unwell**

Causes vary greatly (see table 1<sup>1</sup>), as may the patient's condition, even those with the same cause. Purpura in well patients may be acute, chronic, or recurrent.

**What other features in the history and examination should I consider?****Age**

Differential diagnoses may vary with age, as follows.

**Neonates and infants**

- Moderate to severe congenital bleeding disorders<sup>11 12</sup>
- Acquired thrombocytopenia secondary to sepsis or leukaemia
- Possible child abuse.<sup>11 12</sup>

**Children (in addition to above diagnoses)**

- Immune thrombocytopenic purpura (ITP)
- Vasculitic illnesses such as Henoch-Schönlein purpura (HSP) (fig 3<sup>1</sup>) or viral infection (fig 4<sup>1</sup>)
- Milder congenital bleeding disorders.<sup>11</sup>

**Adults<sup>7</sup>**

- ITP
- Bone marrow failure syndromes:
  - Primary, such as myelodysplasia or leukaemia
  - Secondary, such as malignant bone marrow infiltration
- Nutritional deficiencies
- Medications
- Degenerative diseases such as senile purpura
- Acquired haemophilia
- Mild congenital bleeding disorders may present for the first time.

**Time course**

A short acute illness should raise the suspicion of sepsis. Recent viral illness or immunisation in a well child may precipitate ITP, HSP, or a general vasculitic viral rash presenting as fine petechial haemorrhages.<sup>1 8</sup>

**Distribution of purpura**

In thrombocytopenia the rash is often on the lower limbs and in crying or vomiting children around the head and neck (fig 4<sup>1</sup>). Bruising on the trunk, ears, and face in children, which cannot be adequately explained, is suspicious of non-accidental injury or a severe congenital bleeding disorder.<sup>11</sup>

**Drug history**

This is important, particularly if a new drug has been started, because drug related purpura is well recognised. History of a recent blood transfusion may be relevant as post-transfusion purpura is a rare but serious complication, characterised by severe thrombocytopenia 5-10 days after transfusion.<sup>13</sup>

Table 1<sup>1</sup> outlines clinical findings that may distinguish between causes of purpura.

**Should I refer or investigate?**

Meningococcal sepsis must be excluded in any acutely ill patient with a purpuric or petechial rash; immediate referral to emergency secondary care services is needed.<sup>2</sup> An urgent outpatient referral is not appropriate.<sup>2-5</sup> If invasive meningococcal disease is suspected, administer parenteral antibiotics immediately (benzylpenicillin or cefotaxime), but this should not delay urgent transfer to hospital.<sup>2 3</sup>

In any other patient with a petechial or purpuric rash, severe thrombocytopenia must be excluded urgently. Children and young people should be referred immediately for a full blood count and assessment.<sup>5</sup> A full blood count within 48 hours is recommended in adults, to exclude leukaemia,<sup>5</sup> but may be needed more urgently depending on the clinical assessment. A coagulation screen is also indicated in adults to identify disorders such as acquired haemophilia. ITP, which has an incidence of 2-3/100 000 in adults and children, is an important differential diagnosis.<sup>14</sup>

Advise patients who are not referred immediately to avoid aspirin and non-steroidal anti-inflammatory drugs, which can increase the bleeding diathesis. Ensure the patient's or carer's contact details are available and that the laboratory has 24 hour contact details of the clinician to whom results should be sent. Accurate clinical details also facilitate interpretation of results.

If the full blood count is normal, evaluate and manage the patient according to the most likely underlying diagnosis (table 1<sup>1</sup>).

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### Education into practice

For all blood test requests, ensure clinical details are accurate and that there is a 24 hour contact number for the clinician to whom the results should be sent

### How patients were involved in the creation of this article

This article was submitted before we asked authors to involve patients and report any contributions.

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## Table

**Table 1 | Clinical features of conditions associated with a purpuric or petechial rash†**

Condition	Onset	Other clinical features
<b>Acutely unwell patients</b>		
Acute bacterial sepsis, including invasive meningococcal disease <sup>1-4</sup>	Rapid	<ul style="list-style-type: none"> <li>• Meningism is not always present</li> <li>• Apart from purpura, more specific features include: bulging fontanelle in children under 2 years, altered mental state, unusual skin colour, cold extremities, delayed capillary return, hypotension or shock, stiff neck, seizures, and focal neurological deficits<sup>1-4</sup></li> <li>• Non-specific features include: lethargy, irritability, refusal to eat or drink, nausea or vomiting, headache, fever and muscle aches or joint pain in the extremities and legs, respiratory symptoms or signs, and difficulty breathing<sup>2,3</sup></li> </ul>
Acute leukaemia <sup>5</sup>	Acute to sub-acute	<ul style="list-style-type: none"> <li>• May not be acutely unwell at presentation</li> <li>• May have widespread petechial haemorrhages (fig 2<sup>1</sup>) or ecchymoses on limbs and trunk</li> <li>• Lethargy, pallor, fever</li> <li>• Bony pain or limp: common presentations in children<sup>5</sup></li> <li>• Recurrent infection or sepsis</li> <li>• Lymphadenopathy and hepatosplenomegaly</li> </ul>
<b>All other patients</b>		
Non-accidental injury*	Variable	<ul style="list-style-type: none"> <li>• Consider presence of parental and environmental risk factors</li> <li>• Anomalous or no explanation for findings</li> <li>• Bruising on trunk, ears, or face</li> <li>• Petechiae in SVC area may indicate strangulation</li> </ul>
Idiopathic thrombocytopenic purpura	Acute	<ul style="list-style-type: none"> <li>• All ages, often well, but presentation may be worrying</li> <li>• Purpura can be extensive and affect mucosal membranes (epistaxis, bleeding gums)</li> <li>• Children: recent viral infection or immunisation</li> <li>• May be associated with connective tissue or autoimmune disorders</li> </ul>
Drugs	Variable	<ul style="list-style-type: none"> <li>• Many are reported to cause thrombocytopenia, but fewer cause purpura</li> <li>• Commonly implicated: co-trimoxazole, quinine, carbamazepine, valproate, anticoagulants, antiplatelet agents, steroids in older patients</li> </ul>
Congenital bleeding disorders including haemophilia and von Willebrand disease	Chronic history	<ul style="list-style-type: none"> <li>• Personal or family history of bleeding, especially with dental or surgical challenges</li> <li>• Oral mucosal bleeding, epistaxis or menorrhagia, unexplained bruising</li> </ul>
Acquired haemophilia*	Acute or sub-acute	<ul style="list-style-type: none"> <li>• Secondary immune phenomenon more often in older adults or peri-partum</li> <li>• Often presents with extensive soft tissue bleeding</li> </ul>
Non-leukaemic bone marrow failure (e.g. myelodysplasia, aplastic anaemia, solid tumour infiltration) <sup>7</sup>	Sub-acute	<ul style="list-style-type: none"> <li>• Associated symptoms and signs include lethargy, pallor, recurrent infection and shingles</li> <li>• Symptoms of primary cancer; lymphadenopathy, or hepatosplenomegaly may be present</li> </ul>
Vitamin deficiency**	Sub-acute	<ul style="list-style-type: none"> <li>• B<sub>12</sub> deficiency causing pancytopenia; may be associated with peripheral neuropathy</li> <li>• Folate deficiency causing pancytopenia; consider dietary factors</li> <li>• Vitamin C deficiency causing collagen defect: spongy bleeding gums, painful joints, and corkscrew hairs</li> </ul>
Senile purpura	Chronic	<ul style="list-style-type: none"> <li>• Common in patients &gt;70 years of age; caused by skin fragility and exacerbated by drugs such as aspirin and steroids<sup>7</sup></li> </ul>
Raised SVC pressure	Acute	<ul style="list-style-type: none"> <li>• SVC distribution of petechial haemorrhages including around eyes and mouth</li> <li>• History of severe coughing or vomiting</li> </ul>
Vasculitis	Sub-acute to chronic	<ul style="list-style-type: none"> <li>• Systemic lupus erythematosus: features of connective tissue disease, such as arthralgia and malar rash</li> <li>• Henoch-Schönlein purpura: preceding URTI; purpura usually on shins, buttocks, and posterior thighs (fig 3<sup>1</sup>)</li> <li>• Viral infections: a fine petechial rash with an otherwise benign viral illness, often with coryzal symptoms; usually resolves as the child recovers<sup>1,8</sup></li> </ul>

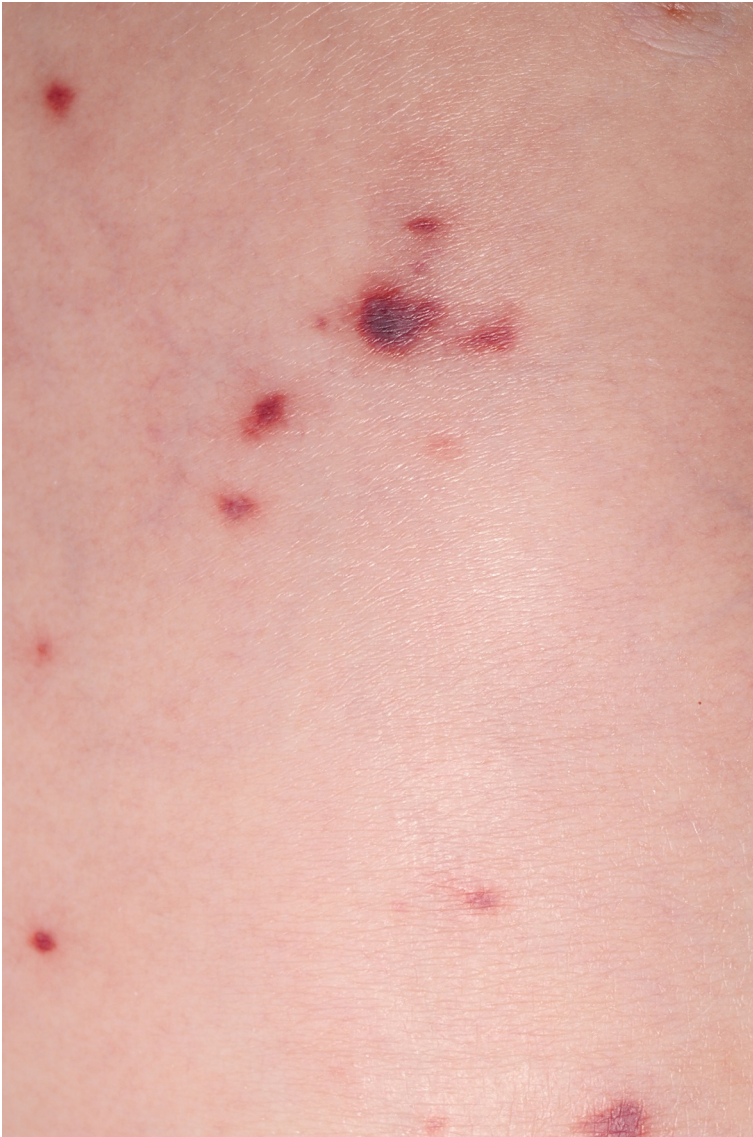
\*Note: some conditions causing purpura, such as child abuse, acquired haemophilia, and vitamin deficiency, may present in both well and unwell patients.

†Abbreviations: SVC=superior vena cava; URTI=upper respiratory tract infection.

‡Onset: rapid=hours; acute=days; sub-acute: weeks to months.

§May also be associated with thrombocytopenia.

## Figures



**Fig 1** Petechial rash in invasive meningococcal disease



**Fig 2** Petechial rash on the forearm of a child presenting with acute lymphoblastic leukaemia



**Fig 3** Vasculitic rash in Henoch-Schönlein purpura



**Fig 4** Fine petechial rash in infant with a viral illness and vomiting