PANS PANDAS UK
awareness support education

STATEMENT on the increase in scarlet fever and invasive group A streptococcus above seasonally expected levels.

PANS PANDAS UK - 6 December 2022

As the only UK charity supporting families managing children with Paediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal Infections (PANDAS), we wish to draw parents', educationalists' and clinicians' attention to key points to be aware of in the management of suspected cases of group A Streptococcus (GAS) and scarlet fever in children who are diagnosed with or suspected to have PANDAS.

GAS can cause an acute onset of neuropsychiatric symptoms related to the immune-mediated condition PANDAS [1], which may arise in the early or later stages of GAS infection [2].

What is PANDAS?

PANDAS is a post-infectious immune-mediated condition triggered by GAS. On average, onset usually happens between 3 and 13 years-of-age. Symptoms often begin incredibly abruptly overnight or rapidly develop over the course of a few weeks. Children with PANDAS display symptoms such as extreme obsessive-compulsive behaviours, motor and vocal tics, severe separation anxiety and behavioural regression. The symptoms can be so severe that many children become housebound. Rage, restricted food intake, sleep disturbances and urinary problems are also frequently observed. Children with this condition may also progress to suffer sensory sensitivities, hallucinations and thoughts of self-harm and suicide.

The latency period between the incident GAS infection and the onset of PANDAS is currently unknown [2] as a) many GAS infections that precede the development of PANDAS are either subclinical or so mild that they do not result in medical attention (as for acute rhematic fever), and b) the common lengthy delays experienced before clinicians identify that a child has experienced an acute onset of neuropsychiatric symptoms triggered by a GAS infection.

Symptoms of scarlet fever and complications of GAS infection

Streptococcus pyogenes (also known as group A Streptococcus [GAS]) can cause the common childhood infection scarlet fever. UK Health Security Agency (HSA) guidance [3] advises that the first symptoms of scarlet fever often include a sore throat, headache, fever, nausea and vomiting. After 12 to 48 hours, a characteristic fine red rash that feels like sandpaper develops. Typically, the rash first appears on the chest and stomach, then rapidly spreads to other parts of the body. On more darkly pigmented skin, the rash may be harder to spot, although the 'sandpaper' feel should be present.

Additional symptoms of scarlet fever include:

- fever over 38.3°C (101°F) or higher is common
- white coating on the tongue which peels a few days later, leaving the tongue looking red and swollen (known as 'strawberry tongue')
- swollen glands in the neck
- feeling tired and unwell
- flushed red face, but pale around the mouth. The flushed face may appear more 'sunburnt' on darker skin
- peeling skin on the fingertips, toes and groin area, as the rash fades

The first symptoms usually appear 2 to 5 days after infection. However, the incubation period may be as short as 1 day and as long as 7 days. People can be infectious for 2–3 weeks after the onset of symptoms, unless they receive treatment [4].

GAS can also cause ear infections, throat abscesses, pharyngitis, impetigo, pneumonia, sinusitis or meningitis. On **rare** occasions, GAS can cause severe disease, including invasive group A streptococcus (iGAS), streptococcal toxic shock syndrome, necrotising fasciitis and septicaemia. Whilst such complications arise in the early stages of GAS infection, subsequent

immune-mediated complications including acute glomerulonephritis, acute rheumatic fever, Sydenham's Chorea and PANDAS can arise later.

Scarlet fever and iGAS are notifiable diseases, which means that all detected cases should be reported to public health authorities. Children who have recently had chickenpox or influenza are more likely to develop a more serious infection during an outbreak of scarlet fever; evidence suggests that chickenpox is the most common risk factor for iGAS disease in children [5].

Clinicians

The UKHSA recommend prompt treatment of scarlet fever with appropriate antibiotics in order to significantly reduce the risk of complications and onward transmission, and to maintain a high degree of clinical suspicion particularly in patients with preceding viral infection (including chickenpox) or close contacts of scarlet fever [6]. NICE advice is that antibiotics should be prescribed promptly, regardless of the severity of illness. NICE also advise that throat swabs and blood tests are not routinely indicated for the diagnosis of scarlet fever [4]. UKHSA advise that a negative throat swab does not exclude the diagnosis [5]. NICE advise that the measurement of serum anti-streptolysin O (ASO) antibody titres are not useful in acute infection, but may be helpful in the diagnosis of post infection complications [4].

Patients diagnosed with PANDAS have already experienced the acute onset of neuropsychiatric symptoms following a GAS infection and may develop an escalation of their neuropsychiatric symptoms if they contract a subsequent infection. This escalation may happen before any physical symptoms become apparent. As with other symptoms of GAS infection, clinicians should consider prescribing appropriate oral antibiotics promptly for suspected GAS infections in children diagnosed with or suspected to have PANDAS, regardless of the severity of illness, such as phenoxymethylpenicillin for 10 days as a first-line treatment [4]. Follow up should be arranged if symptoms worsen or have not improved after 7 days.

Household contacts of scarlet fever cases have been found to have a low but increased risk of iGAS disease in the 2 months after the onset of scarlet fever [5,7]. Clinicians should advise patients and their parents/guardians to be vigilant for any symptoms which might suggest these complications and to seek medical help immediately if concerned.

Parents/Caregivers of Children with PANDAS

Many parents are highly concerned about the reports in the media regarding iGAS and the heart-breaking cases of the children who have sadly died. It is important to be familiar with the symptoms of scarlet fever (see above) and if you are concerned for any reason about your child's health, please seek appropriate medical assistance immediately.

If you are concerned about your child's vulnerability to an escalation of PANDAS symptoms, please consult your GP/consultant for advice. It may also be helpful to ensure that your child's education setting understands that you are concerned about the additional potential risk of a GAS infection to your child given their PANDAS diagnosis or suspected PANDAS diagnosis. You may want to share this statement with your child's GP and education setting.

The usual treatment for scarlet fever is a 10-day course of antibiotics. Any fever present will usually subside within 24 hours of starting antibiotics; it is important to take the entire course of treatment to completely clear the infection and to protect other individuals from becoming infected [3].

Education Settings

Schools, nurseries and other childcare settings should promptly notify their local Health Protection Team (HPT) of a suspected scarlet fever outbreak. An outbreak of scarlet fever is defined as a credible report of two or more probable or confirmed cases of scarlet fever among children attending the same school/nursery or other childcare setting that are notified within 10 days of each other and have an epidemiological link; for example, cases among children in the same class or year group [5].

Parents should be encouraged to take children who may have scarlet fever to their GP for a clinical diagnosis and appropriate testing. Children or nursery/school staff who refuse treatment with antibiotics should be excluded from the educational setting until their symptoms completely resolve [5]. An untreated infection increases the risk of complications such as acute rheumatic fever and can lead to long-term carriage of GAS.

As recommended in the current national 'Guidance on Infection Control in Schools and other Child Care Settings' [5], staff and parents should be reminded that children and adults with scarlet fever should not return to nursery or school until at least 24 hours after starting treatment with an appropriate antibiotic. It is important to take the entire course of treatment to completely clear the infection and to protect other individuals from becoming infected [3]. Good hygiene practices, such as hand washing, remain the most important step in preventing and controlling the spread of infection [5].

About PANS PANDAS UK

PANS PANDAS UK is the only UK charity supporting children and families living with these conditions. We provide information, advocacy and community support to patients and carers. We raise awareness of the symptoms and treatment options for healthcare professionals so that they are better equipped to recognise when a child may have PANS or PANDAS. We are working to provide training in educational settings so that families and children living with these conditions receive the support they so desperately need.

References

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