with nocturnal hypoventilation (daytime drowsiness, headache, poor sleep pattern), then overnight respiratory monitoring is required and BIPAP (bi-level positive airway pressure) intervention should be considered.

Schooling and independence. It is hard for a child to lose ambulation and to see his peers/siblings outpace him. All attempts must be made to respect the individual through appropriate school placement, an electric wheelchair and access to computer skills. Children with DMD are often in mainstream education and may have to transfer to a special school once they are no longer ambulant.

Ethics and counselling. Older children may have better insight into their condition than parents realise. However, their concerns tend to focus more on how the disability limits their activities of daily living. Thoughts of their longevity are not an immediate priority, while this dominates for many parents. Open-ended questions enable the child to respond if they have issues, but a child should not be confronted with a list of health concerns. Routine screening of unaffected/possibly pre-symptomatic male siblings is not recommended. Refer the family for formal genetic counselling to ensure they fully understand the implications of testing and the reliability of such screening. If maternal aunts or the mother requests screening and if there is DNA confirmation, then linkage analysis may enable carrier status to be established. Otherwise CK levels sometimes, but not reliably, indicate whether they are carriers.

Parents and carers. Their needs are often forgotten or there is no adequate support network. Encouraging them to contact the Muscular Dystrophy Foundation (http://www.mdsa.org.za/index.htm) may help them to cope with the emotional and physical burdens.

The future

There is currently no cure for DMD. Gene therapy may be one intervention, but to date it is not available. Other possible therapies include the role of PTC124, an agent which allows 'read through' of a premature stop codon and the production of dystrophin. Such treatments are targeted at specific genetic mutations. It is important to ensure that the DNA from all affected children is stored. The expanding capacity of genetic screens is resulting in further mutations being identified in children previously reported deletion negative. Some 20 years ago children with DMD died a difficult death – often by 14 years of age. Currently, with a multidisciplinary approach, children can survive with a good quality of life to over 30 years of age. Management is not directed at prolongation of life, but more at the rights of the individual to reach their full potential.

Summary

- DMD affects mainly boys.
- The clinical onset is typically by 3 - 5 years of age.
- The creatine kinase is usually >10 000 mmol/l.
- Confirming the diagnosis is essential, as the management and inheritance differ from other muscular dystrophies.
- Prophylactic corticosteroids and ACE inhibitors are known to improve survival and quality of life.
- Multidisciplinary care results in better quality of life and longevity into the second or third decade.

References available at www.cmej.org.za

Post-streptococcal neuropsychiatric movement disorders or Sydenham's chorea spectrum disorder: an update on management

KATHLEEN G WALKER, *MB CHB, DCH*

Medical Officer, Division Paediatric Neurology, Paediatric Cardiology and Kidzpositive HIV Clinic, Red Cross War Memorial Children's Hospital and Groote Schuur Hospital, University of Cape Town

JO M WILMSHURST, *MD*

Paediatric Neurologist, Red Cross War Memorial Children's Hospital, Cape Town

Correspondence to: Kathleen Walker (buley@iafrica.com)

Sydenham's chorea (SC) was first described in 1686 by Thomas Sydenham.[1] It is a major criterion for the diagnosis of acute rheumatic fever and its presence alone is sufficient to make this diagnosis.[2] Up to 60% of people who present with SC will later develop rheumatic heart disease.[3]

SC is an anti-neuronal, antibody mediated neuropsychiatric dis-order.[4] Antibodies that arise in response to group A beta-haemolytic streptococcus (GABAS) infection cross-react with epitopes on neurons within the basal ganglia, frontal cortex and other regions.[4] A cerebral arteritis with cellular degeneration occurs.[5] This results in dopaminergic dysfunction, which has an effect on movements, attention and emotion.[6]

Clinical presentation

The clinical features of SC include both neurological abnormalities and psychiatric disorders. The former comprise involuntary choreatic movements, voluntary movement incoordination, muscular weakness and hypotonia.[7] Psychiatric disorders include emotional lability, hyperactivity, distractibility, obsessions and compulsions.[2] Difficulty in the execution of activities of daily living results, such that the condition impacts negatively on the quality of life of affected children.

Choreatic movements are involuntary, irregular, purposeless, non-rhythmic, abrupt, rapid and unsustained.[8] Movements disappear with sleep and rest. Voluntary movements make the chorea worse.[9] Hypotonia and weakness range from mild to severe. The severe form is termed chorea mollis or chorea paralytica and may be confused with the clinical appearance of a stroke.[1] Obsessions may include harm to loved ones, separation anxiety and fear of contamination, resulting in compulsive washing.[10] Children may have severe chorea (ballistic movements) and/or hypotonia with few psychiatric symptoms or mild chorea with pronounced psychiatric symptoms.[2] A change in behaviour may precede the chorea.

Classic descriptions of SC indicate that it is benign and self-limiting.[11] At best the condition lasts for 6 months, but more usually it has a relapsing course for up to 2 years.[12] It may evolve into a chronic movement disorder.[12] It is important to quantify the severity of symptoms as a therapeutic index. Aron's clinical classification refers to 'mild' in the presence of minimal movements, 'moderate' in the presence of movements of obvious inconvenience to the patient but which do not interfere with self-care, and 'severe' if there are movements sufficiently incapacitating for the patient to require assistance for the activities of daily living.[5]

Therapeutic interventions

Treatment has four main tenets: elimination of the streptococcus, symptomatic treatment of the involuntary movements, incoordination and psychiatric symptoms, treatment of the immune and inflammatory response and supportive measures.

Primary treatment: elimination of streptococcus

Treatment with penicillin is mandatory to eliminate the streptococcus. When SC is diagnosed penicillin 500 mg twice daily for 10 days should be given, and rest is advised.[13] Adverse outcomes and a chronic relapsing course of SC are more common in children who do not receive 10 days of penicillin and bed rest.[12]

Prophylaxis with long-term penicillin is primarily given to protect the heart. Intramuscular benzylpenicillin every 28 days or oral penicillinVK 250 mg twice daily is advocated as secondary prevention of rheumatic heart disease.[13] Patients must also be advised to seek primary treatment for future streptococcal sore throats.

Symptomatic treatment

There is no globally accepted protocol to treat SC. Agents that affect the neurotransmitters dopamine and GABA are used as symptomatic treatment. Dopamine receptor antagonists include haloperidol, which is an effective symptomatic medication but which must be titrated slowly to reach maximum effect with minimal toxic manifestations.

In South Africa haloperidol is first-line therapy as it is cost effective and readily available. Use of haloperidol should be based on a regimen of 'start low and go slow'; 0.025 mg/kg/day in divided doses going up to a maximum of 0.05 mg/kg/day in divided doses.[12]

GABA is a neurotransmitter which inhibits dopaminergic over-activity.[8] Sodium valproate enhances the action on GABA; hence it is sometimes used to treat chorea.[8] Recommended doses vary between 15 mg/kg/day to 20 mg/kg/day in 2 or 3 divided doses. Sodium valproate has been recommended as treatment where trials with haloperidol have failed and the chorea remains severe.[11] Phenobarbitone is not recommended for symptomatic treatment.

There are detailed descriptions of the psychiatric manifestations in SC but no reports of treatments specifically targeting these symptoms.

Therapeutic interventions: immunological treatments

Antibodies reactive with neuronal tissue in the serum of patients with SC demonstrate that the condition is a humorally mediated autoimmune condition.[10] Imunomodulatory therapies are described using corticosteroids, intravenous immunoglobulins (IVIG) and plasma exchange.[14] Controlled studies are needed to definitively clarify efficacy of these interventions.

Supportive measures

Management of SC requires a multi-disciplinary approach. Management of co-morbid psychopathologies is important. Supportive psychotherapy and family therapy are recommended. Regular attendance should be encouraged for follow-up assessments and to receive benzathine benzylpenicillin injections. Helping educators to understand the condition will promote tolerance and understanding and help them to ensure that children with SC are exposed to minimal teasing and bullying.

Every child with SC must be notified and must undergo echocardiography.

Conclusion

SC has a clinical spectrum from severe chorea with mild psychiatric symptoms to minimal chorea with severe psychiatric symptoms. It is better to refer to SC as a post-streptococcal, autoimmune, neuropsychiatric movement disorder, which encompasses all aspects of the disease.

As SC is synonymous with acute rheumatic fever it must always be notified. The main goal of treatment should be primary prevention, which requires eradication of poverty and poor living conditions. Until this millennium goal is achieved strategies should target management which reduces the burden of disease in children living with SC.

References available at www.cmej.org.za

Approach to a single granuloma on CT scan

REGAN SOLOMONS, *MB ChB, DCH (SA), MMed (Paed), Cert Paed Neur (SA)*
Consultant Paediatric Neurologist, Tygerberg Children's Hospital, Department of Paediatrics and Child Health, Faculty of Health Sciences, Stellenbosch University

JOHAN SCHOEMAN, *MB ChB, MMed (Paed), FCPaed (SA), MD*
Professor and Head, Paediatric Neurology, Tygerberg Children's Hospital, Department of Paediatrics and Child Health, Faculty of Health Sciences, Stellenbosch University

RONALD VAN TOORN, *MB ChB, MRCPCH (UK), FCPaed (SA), Cert Paed Neur (SA)*
Consultant Paediatric Neurologist, Tygerberg Children's Hospital, Department of Paediatrics and Child Health, Faculty of Health Sciences, Stellenbosch University

Correspondence to: R Solomons (regan@sun.ac.za)

The increasing availability and use of neuro-imaging in children with new-onset partial seizures has resulted in increased identification of intracranial granulomas.

Neurocysticercosis is the most common parasitic infection of the central nervous system and a major cause of seizures in resource-poor countries.[1] Humans develop neurocysticercosis when they accidentally ingest eggs of the pork tapeworm. It is not related to eating undercooked pork and therefore may occur in vegetarians and persons of religions that prohibit the eating of pork. In South Africa, neurocysticercosis is the most common cause of granulomas detected by computed tomography (CT) scanning. The second most common cause of intracranial granuloma is tuberculoma, which may occur in isolation or in association with tuberculous meningitis. Lymphoma and toxoplasmosis should be considered as possible causes for intracranial ring-enhancing lesions in the immune-compromised child.

Radiological differentiation between a tuberculoma and a single cysticercal granuloma is difficult.[2] Correct diagnosis is important because misdiagnosis of a tuberculoma in a patient with neurocysticercosis can result in 6 months' unnecessary high-dose antituberculosis therapy. Similarly, not identifying tuberculosis in a patient could be disastrous. We present an approach to the diagnosis and management of a single granuloma.

Clinical features

Clinically, children with neurocysticercosis often present with seizures. Headaches, altered level of consciousness, focal neurological signs and cerebellar signs present less frequently. If a tuberculous origin is suspected, the following clinical information is informative:

- positive tuberculosis contact
- features of systemic tuberculosis, including failure to thrive, night sweats and coughing for longer than 2 weeks
- focal neurological signs.

Exclusion of active tuberculosis is a priority and has the most impact on patient outcome and management.

Diagnosis

The radiological appearance of neurocysticercosis may range from non-enhancing cysts (viable cyst with no host immune response) to enhancing cysts or granulomas (degenerating cyst with surrounding immune response) to a calcified lesion (dead cyst with total resolution).[3] Contrast CT imaging is adequate in most cases of suspected neurocysticercosis. CT is superior for the detection of calcified granulomas, while magnetic resonance imaging (MRI) is better for visualising parasitic cysts. In a minority of cases, a scolex (small, eccentric internal speck of enhancement or calcification within the cyst) may be visualised. This finding is pathognomonic of neurocysticercosis (Fig. 1).

The second most common CT-enhancing lesion is a tuberculoma (Fig. 2). Imaging features favouring a diagnosis of tuberculoma rather than parasitic granuloma include:

- size >2 cm
- lobulated, irregular shape
- presence of marked oedema
- persistent, focal neurological deficit.

Because of the difficulty of differentiating between a tuberculoma and cysticercus