Scimitar syndrome as a differential diagnosis in a child with recurrent wheeze

Respiratory symptoms of cough, wheeze, and breathlessness account for 40% of referrals to a general paediatric clinic. The majority of these children suffer from “wheeze secondary to upper respiratory tract infection” and “asthma.”

A 7 year old girl was referred by her general practitioner to the clinic with a two month history of persistent cough and recurrent wheeze; she had been treated for suspected asthma with fluticasone and salbutamol since early childhood. There was a history of infantile eczema. She was growing well on the 50th centile. General examination was normal. There was no cyanosis or clubbing. Respiratory and cardiovascular system examinations were unremarkable.

She had been admitted at the age of 14 months with cough and wheeze; chest x ray showed right lower lobe consolidation which improved on antibiotics. Both radiographs were studied and the pattern of curvilinear density (scimitar) in the right lower zone suggested scimitar syndrome was recognised. She was referred to the paediatric cardiology department for echocardiography, which showed dilated right atrium, right ventricle, and a branch of the right pulmonary vein draining into the inferior vena cava, a mild variant of scimitar syndrome.

A cardiac catheterisation and coil embolisation of the systemic pulmonary collateral from the descending aorta to the right lower lobe is scheduled.

Scimitar syndrome is a name given to the triad of: (1) curvilinear vascular density in the right lower zone; (2) hypoplastic right lung; and (3) dextroposition of the heart. It has a wide spectrum of presentation and may sometimes only present in adulthood with symptoms of wheeze, recurrent chest infections, or pulmonary hypertension.

It remains a notoriously difficult diagnosis to make without a strong index of suspicion. In this case, pattern recognition on chest radiograph helped us to suspect the diagnosis. Examination and ECG may be entirely normal or just show right sided strain. Echocardiogram may also be normal or show dilated right sided chambers (as in this case). Diagnosis can be missed in up to 33% cases by echocardiography. More sensitive tests would include computed tomography scan, cardiac catheterisation, and magnetic resonance imaging with 3D MRA. Obstructive and early symptomatic types will usually need corrective surgery after stabilisation. Milder scimitar variant will probably do well with occlusion of the collateral supply.

We have presented this case to highlight the fact that one has to keep an open mind regarding the final diagnosis in any child with recurrent wheeze, as all wheezes are not “asthma.”

P R Desai, M Babu
Department of Paediatrics, St John’s Hospital, Chelmsford CM2 9B, UK; prpravin@yahoo.com

References

Assessment of acute admissions by middle grade trainees and consultants will reduce the need for overnight hospital admissions

We carried out an audit to assess the impact on hospital admissions of patients being assessed by either middle grade trainees (residents) or consultants in a district general hospital (DGH). Our aims were to establish:

- Number of children kept in hospital overnight and those discharged the same day.
- Number of readmissions of those discharged the same day.
- Any adverse events in those discharged home the same day.

We studied retrospectively all acute admissions to the children’s wards at Doncaster Royal Infirmary, a medium sized district general hospital, over the months of January and July 1998. We excluded all surgical and non-acute admissions. At the time of the study the Children’s Hospital did not have a day or acute assessment unit. Therefore the children were reviewed following admission to the wards. Whether trainees or consultants reviewed patients was an entirely random process, dependent on willingness and time to carry out ward rounds in late afternoon or early evening. The review could also be triggered by nursing staff or parents. The interval between the time of admission to the ward and the time the patients were reviewed varied from immediate review to a few hours. The decision to discharge children was usually taken jointly by medical and nursing staff, provided that parents were willing to look after their children at home. The parents of children discharged home on the same day as admission were given open access to the children’s ward—that is, they could either telephone the ward for advice or return with the child if concerned.

A total of 512 sets of case notes were reviewed by MMM and RAS. A pro forma was used to collect the data, which was stored on an Excel spreadsheet.

A total of 173 (34%) patients were under 1 year, 150 (29%) were 1–2 years, 53 (10%) were 3–4 years, 41 (8%) were 5–6 years, and 95 (18%) were over 6 years (fig 1). The source of referral was documented in 499 case notes. Of these, 287 (58%) were via a general practitioner, 178 (36%) were via the accident and emergency department, and 29 were from other sources. The commonest reason for admission was breathing difficulties followed by fever.

Of the 512 patients admitted, 260 (51%) were reviewed by middle grade trainees or consultants. Of those reviewed, 109 (42%)
were discharged home the same day. The age
group distribution (fig 1) and reason for
admission (fig 2) of those reviewed was simi-
lar to that of the total sample. More children
under 1 year were kept in overnight than were
discharged home the same day; the reverse
was true for those in the 1–2 year and 3–4 year
age groups. The reason for admission of those
discharged home the same day was also simi-
lar to that of the total sample. Slightly more
patients were admitted in January than in July;
but more patients were discharged home the
same day in July than in January (26% v
15%; fig 3). This could well be due to the fact
that there is more pressure on beds in the
winter months. However, it could also be due
to a different spectrum and severity of
diseases.

Of those discharged home the same day,
seven (6%) were readmitted within seven
days, four because of the same complaint and
three with a different complaint. There were
no adverse events. Those who were reviewed
but kept in overnight had a similar distribu-
tion of the reason for admission to that of the
total sample and those who were reviewed but
with an excess of vomiting and/or diarrhoea.

In conclusion, assessing the need for
admission resulted in 20% of all admissions
(40% of those reviewed) being discharged
home the same day. Vomiting and/or diar-
rhoea were more likely to result in patients
being kept in overnight. We believe the
number of patients who can be discharged
home the same day will be much higher if all
acute admissions are reviewed and assessed
in the way described. This policy seems safe
and acceptable to parents.

With the planned reduction in the number
of specialist registrars, it seems that expand-
ing the number of consultants would achieve
the dual benefit of moving closer towards a
consultant provided service and will also lead
to reduction in the number of children requir-
ing an overnight hospital admission.

Parkinson-like syndrome as the major presenting symptom of
Epstein--Barr virus encephalitis

The main symptoms of Epstein--Barr virus
encephalitis (EBV) encephalitis are fever,
seizure, bizarre behaviour, headache, and
metamorphosis.1 Bradykininesia, akathisia, in-
voluntary hand movements, drooling, and
torticollis are symptoms of Parkinson-like
syndrome, which has never been described as
a manifestation of EBV encephalitis. We
report the case of a previously healthy boy
who presented with Parkinson-like syndrome
as the major symptom of EBV encephalitis.

A 12 year old, previously healthy boy was
referred to our hospital because of severe
cough with sputum and intermittent fever for
seven days. Abdominal discomfort and vomit-
ing were also noted one day before admission.
On admission, his consciousness was clear
without focal neurological sign, no hepato-
splenomegaly, no lymphoadenopathy, and no
exudative tonsillitis or skin rash. There was no
previous personal or family history of seizure
disorder or migraine, and both the boy and his
family denied being exposed to some possible
hallucinogenic or neuroleptic drug. Blood
analysis was normal except for a mild
leucocytosis with a left shift (10 500/μl, 84.6%
neutrophils), and there were no atypical
lymphocytes. C reactive protein (CRP)
level was 0.17 mg/dl (normal <0.3 mg/dl).

After admission, mucocutaneous agents
and bronchodilators were prescribed. Fever, ab-
dominal discomfort, and vomiting subsided
soon. Twenty four hours after admission, his
condition deteriorated with drowsiness, invol-
tuntary rhythmic finger tapping movement,
resting tremor, bradykinesia, photophobia, and
staring eyes. But perceptual distortion was not
noted. Cerebrospinal fluid (CSF) examination yielded yellow, clear fluid and
normal opening pressure without microor-
organisms on Gram stain or culture. The cell
count, protein, and sugar of CSF were all
within normal limits. Table 1 lists laboratory
evaluations for EBV and other possible patho-
gens of encephalitis. Brain magnetic reso-
nance imaging (MRI) showed no abnormali-
ties. Tc-99m HMPAO brain SPECT (Tc-99m
hexamethylpropyleneamine oxime single
photon emission tomography) showed dimin-
ished perfusion in the region of the right cau-
date nucleus. Electroencephalography (EEG)
revealed diffuse slowing of background activ-
ity. His signs and symptoms showed gradual
improvement under close observation in the
following three weeks. He was then dis-
charged in a stable condition. Follow up four
months later showed no residual neurological
sequelae.

Parkinson-like syndrome (extrapyramidal
symptoms) is characterised by various neuro-
logical symptoms including akathisia, brady-
kiniesia, torticollis, drooling, and involuntary
hand movement. This syndrome develops in
at least a quarter of children treated with
neuroleptics due to disruption of the balance
between the dopaminergic system and the
cholinergic system within the basal ganglia.
But Parkinson-like syndrome has also been
recognised as a sequel of acute viral encepha-
litis, including coxsackie B, cytomegalovirus,
measles, herpes simplex virus, Japanese B
encephalitis virus, and encephalitis lethar-
gica. Mycoplasma pneumoniae infection has also
been recognised as a cause of Parkinson-like
syndrome.1 In our patient, exposure to possi-
ble hallucinogenic or neuroleptic drugs was
denied. Serological tests and culture for other
possible pathogens were negative. EBV ence-
phalitis was diagnosed by serological and
CSF polymerase chain reaction findings
which fulfilled the diagnostic criteria.

EBV encephalitis is generally considered to
be a benign, self limited disease associated
with few sequelae. However, an incidence of

<table>
<thead>
<tr>
<th>Day 1</th>
<th>Day 14</th>
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<tbody>
<tr>
<td>EBV VCA IgM</td>
<td>1/32 [positive]</td>
</tr>
<tr>
<td>EBV VCA IgG</td>
<td>1/160</td>
</tr>
<tr>
<td>EBV determined nuclear antigens</td>
<td>Negative</td>
</tr>
<tr>
<td>EBV CSF PCR</td>
<td>Positive</td>
</tr>
<tr>
<td>Mycoplasma pneumoniae IgM</td>
<td>&lt;1/40</td>
</tr>
<tr>
<td>Culture for enterovirus</td>
<td>Negative</td>
</tr>
<tr>
<td>Coxsackie B virus IgM</td>
<td>Negative</td>
</tr>
<tr>
<td>Cytomegalovirus IgM</td>
<td>Negative</td>
</tr>
<tr>
<td>Herpes simplex IgG and IgM</td>
<td>Negative</td>
</tr>
<tr>
<td>JBE virus IgM</td>
<td>Negative</td>
</tr>
</tbody>
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neurological sequelae after EBV encephalitis as high as 36% was reported by Domachowske and colleagues. The present case, therefore, not only draws attention to the role of EBV in infectious neurological disorders, but also suggests that an EBV aetiology should be considered in cases of Parkinson-like syndrome in childhood.

J-C Hsieh, K-H Lue, Y-L Lee
Chung Shan Medical University Hospital, 23 Sec. 1 Taijunggang road, Shi Chiu, Taichung, Ta, Republic of China; kent0837@yahoo.com.tw

References

Hyperextension of spine: unusual presentation of Guillain-Barré syndrome

Guillain-Barré syndrome (GBS) classically presents as ascending symmetric areflexic weakness with positive sensory symptoms. Recently, we managed a child presenting with unusual posture and hyperextension of the whole spine.

A 9 year boy presented with inability to hold books and write, and a limp. Over 12 hours he had developed tingling sensations and pain in the calf muscles; pain progressed to involve the neck and back by 24 hours and he was unable to flex his neck and extend his limbs. During the next two days weakness increased, especially of the lower limbs. By day 4, he had developed hyperextension of the cervical and thoracolumbar spine with flexed and adducted limbs. On day 6 when he presented to us, vital signs including blood pressure were normal and remained so during the hospital stay. He had painful restriction of passive extension at all joints; motor power and tone could not therefore be assessed. He had bilateral symmetrical weakness: shoulders (abductor, adductor, 2/5), elbows (flexor 3/5, extensor 2/5), wrists (dorsiflexor, palmar-flexor, 2/5), finger flexors (2/5), hand grip (20–30%), hip flexors (2/5), knees (flexor, extensors, 2/5), ankle (0/5), toes (0/5). Deep tendon reflexes were absent except for the biceps, which also disappeared by day 12. Cremastic and abdominal reflexes were present; plantars were absent bilaterally. The spine was normal except for hyperextension of the cervical and thoracolumbar region. Respiratory muscles, higher mental functions, speech, cranial nerves, and bowel and bladder functions were normal. A plain radiograph of the spine showed mild thoracic lordosis. Cerebrospinal fluid examination on day 11 showed high protein (95 mg/dl). On day 12, spinal hyperextension and abnormal limb postures disappeared following improvement in pain as a result of analgesic therapy. Kernig’s and Brudzinski’s signs could be elicited. Lasegue’s sign and the straight leg raising test were also positive. Symmetrical hypotonia became obvious.

GBS was suspected in view of progressive bilateral symmetrical weakness, severe radiculopathy, and albumino-cytological dissociation. Nerve conduction studies, performed on day 23, showed reduced nerve conduction velocity in the motor nerves. Sensory nerves were normal. We could not determine if wave conduction velocity. The pain and tenderness subsided gradually. With regular physiotherapy the neck became soft, motor power improved, and he was able to sit unsupported by day 20. Four months later, neurological examination was normal. A stool culture for poliovirus was negative.

Children with GBS frequently have pain in the back and lower limbs, which is aggravated on the straight leg raising test in most of them. These pain syndromes are attributable to radiculitis, an early and predominant feature in GBS. Prominent radiculitis in this case might have led to severe pain in the back, causing generalised paravertebral muscle spasm. This resulted in the unusual posture of hyperextension of the spine.

A K Baranwal, S C Singh
Department of Pediatrics, BP Koirala Institute of Health Sciences, Dharan, Nepal; baranwal1970@yahoo.com

REFERENCES

CORRECTIONS
An error occurred in the letter by S Ashraf and M Z Mughal in the September issue (Arch Dis Child 2002; 87: 263–4). In the fifth paragraph, the first sentence should read “According to the 1991 census data there were approximately 4000, 6–36 month old children of ethnic minority background resident in the city of Manchester.” The journal apologises for the error.

The photograph of children in the Morigate area in Delhi on the cover of the August issue was taken by the American photographer Mark Juerkens of Dallas, Texas.
Intravenous atropine treatment in infantile hypertrophic pyloric stenosis

Hypertrophic pyloric stenosis of infancy is a disorder of early infancy with typical clinical features and well-established radiological appearance. The pyloric canal. Many studies with surgical and medical treatment have been reported over the past fifty years. Pylorotomy has tended to become the favoured method of treatment as with expert paediatric, surgical, anaesthetic, and nursing services and specialised accommodation for infants, the outcome is good with low mortality, short stay in hospital and few complications. However, a variety of studies of medical treatment with anticholinergic drugs and successful outcomes in some large series of cases have also been reported from Sweden, United States of America and the United Kingdom.

Since 1996 this group of workers from Osaka, Japan, has revived an interest in medical treatment with reports of a new regime using methyl atropine nitrate intravenously. To achieve satisfactory short term outcomes considerable variation in drug dosage and modified feeding regimes were necessary which involved much medical supervision and careful monitoring for toxic effects of the drug, which were minimal. The treatment was successful in the relatively small number of infants in the trial (19) with two infants being referred for pylorotomy, no mortality and no serious complications. An interesting part of this paper is the long term clinical follow up of the successfully treated infants over two years and ultrasonography of the pyloric canal which demonstrated the changes in muscle thickness and length of the canal. The disadvantages of the treatment mentioned by the authors are length of stay in hospital and the necessity to continue atropine medication orally after discharge home.

Comparing the use of this anticholinergic drug intravenously with oral treatment using methyl scopolamine nitrate and similar restricted feeding regime, oral methyl scopolamine nitrate suppressed vomiting more quickly and reliably, was also available for subcutaneous injection if vomiting recurred as size of feeds was increased, and no toxic effects were seen in any dosage used. It would be interesting if these workers would be prepared to try the use of methyl scopolamine nitrate intravenously as pharmacologically this compound was reported to have a spasmylic effect on gut two to three times greater than methyl atropine nitrate with lesser central nervous effects.

This paper serves to emphasise once more that these infants should always be treated in paediatric centres where there is a high level of experienced paediatric care and nurses trained for neonatal special care.

**Author’s reply**

We appreciate the interest shown by Dr Beryl Corner with regard to our article.1 Unfortunately, intravenous atropine therapy is not widely accepted in European countries or the United States; it is however now becoming popular in Japan.

We are truly honoured to receive the comments of Dr Corner, who is a pioneering neonatologist and reported medical treatment with methyl scopolamine nitrate for infantile hypertrophic pyloric stenosis (IHPS) in 1955.2 She pointed out that methyl scopolamine might be better than atropine sulfate in terms of effectiveness and side effects. One of the reasons why atropine was used in our study is that methyl scopolamine is not available in our country. Scopolamine butylbromide is an available quaternary ammonium derivative of scopolamine and lacks toxic side effects. However, this agent tastes bitter and is difficult to give orally to infants. Therefore, this agent is only given intravenously in infants with IHPS.

We do not know if it is worthwhile to attempt combination therapy with intravenous scopolamine butylbromide and oral atropine rather than the intravenous and oral atropine therapy. Secondly, we already knew that an intravenous atropine injection of 0.01 mg/kg was effective enough to abolish transiently the phasic and tonic pyloric contractions characteristics of IHPS.3 We used an intravenous atropine injection of 0.01 mg/kg in our study to confirm that those pyloric contractions were the cause of disturbed transpyloric flow in this condition by seeing that their inhibition with the dose of atropine ameliorated symptoms.

We agree with Dr Corner’s last comment, but believe that intravenous atropine therapy is possible not only in high level paediatric centres, but also in general hospitals where infusion therapy with intravenous atropine injections can be done safely in small infants. Clinical trials are now ongoing to establish more efficient treatment strategy for IHPS with medical and surgical therapy in our country.

**H Kawahara**

Consultant Paediatric Surgeon, Osaka Medical Centre and Research Institute for Maternal and Child Health; kawahara@ped.surg.med.osaka-u.ac.jp

**References**


**Hypothermia in a child secondary to ibuprofen**

A 7 year old girl was admitted with right lower lobe pneumonia. On admission her temperature was 39.7°C. After five hours she received ibuprofen (6 mg/kg). Subsequent to this single dose her temperature decreased to 33.5°C (core temperature 34.9°C) over four hours.

On examination her pulse was 90/min, blood pressure 90/50 mm Hg, SaO2 96% in air, and respiratory rate 20/min. Respiratory examination was consistent with signs of right lower lobe consolidation. The rest of the examination, including the central nervous system, was unremarkable.

Results of investigations included: Hb 125 g/l; white blood cell count 10.7 × 10⁹/ℓ platelet count 81 × 10⁹/ℓ; C reactive protein 180 mg/l; blood glucose 4.6 mmol/l. Electrolytes and all other biochemical investigations were normal. Thyroid and cortisol assays were normal. Results of all tests to determine possible bacterial or viral aetiology were all negative (blood and urine culture, viral serology, and tests for mycoplasma). Magnetic resonance imaging (MRI) of the brain was normal.

The hypothermia was so marked that we had to use a hot air spacer blanket to raise her temperature. Despite all the efforts she remained persistently hypothermic for four days (see fig 1).

**Figure 1** Temperature chart. After administration of ibuorifen, the temperature dropped considerably and remained low for five days.
A single dose of hydrocortisone and an albumin infusion were given initially. She was subsequently treated with warmed intravenous fluids for three days and antibiotics for 10 days. She recovered completely and continues to enjoy good health.  

Disordered autonomic function, including cardiorespiratory control, has been suggested to be involved in SIDS. Among their siblings, five of 126 had died of SIDS. All five children were full term infants. The average maternal age, birth weight, and age at death were respectively 27.4 (3.5) years, 3.3 (0.3) kg, and 3.5 (1.1) months. The rates of SIDS in siblings of children with VO were compared to those in the general population using the standardised incidence ratio (SIR), which is the ratio of the observed number to the expected number of cases of SIDS calculated by French incidence rates. The expected number of SIDS was 0.17 and hence the SIR was 29.4 (95% CI 9.5 to 68.6; p < 0.000011). Our result showed an overall significant excess of SIDS among siblings of children with VO. We suspect that recruited children had not come to the centre because of a family history of SIDS. Since children with a positive family history of SIDS could be followed up more regularly than others, we estimated the SIR separately among siblings of children recruited during their follow up and those of children recruited during their first visit, and verified that there was no significant difference in SIR between these cases. Despite the marked decline in SIDS, it is still the leading cause of postneonatal mortality. Better knowledge of other risk factors may allow identification of populations at higher risk and hence in infant mortality from SIDS through the implementation of appropriate prevention measures. Our findings suggest that VO may be involved in SIDS and that children with VO or a family history of VO may be a population at potential high risk of SIDS.

References


Vagal overactivity: a risk factor of sudden infant death syndrome?

Since early 1990, the incidence of sudden infant death syndrome (SIDS) has dropped sharply because of public health campaigns decrying the dangers of the prone sleep position. The other known risk factors, such as preterm birth and young maternal age, are less susceptible to prevention campaigns. Disordered autonomic function, including cardiorespiratory control, has been suggested to be involved in SIDS. Vagal overactivity (VO), characterised by breath holding spells and repeated syncope in specific circumstances, has been described as a manifestation of autonomic dysfunction. To investigate a possible relation between VO and SIDS, we investigated 65 children presenting documented VO; for example, clinical characteristics and a positive test for eye ball compression and/or electrocardiographic monitoring. Parents of these children were interviewed about their family history, especially with respect to the occurrence of SIDS among their other children.

Perforated duodenal ulcer disclosing medium chain acyl-CoA dehydrogenase deficiency

Medium chain acyl-CoA dehydrogenase deficiency (MCADD; McKusiek 201450) typically presents in the first two years of life with recurrent episodes of hypoketotic hypoglycaemia, lethargy, coma, or sudden infant death. The trigger may be fasting, intercurrent infections, anaesthesia, or surgery. Incidence in the UK is estimated at 0.45–1/10 000 live births.1 We describe the case of a child who presented with marked encephalopathy unexplained by perforated duodenal ulcer, which led to the diagnosis of MCADD.

A 2 year old girl presented with a three week history of corzyl symptoms and three day history of frequent coffee ground vomitting. She was shocked, and had hepatomegaly and decreased conscious level. Blood glucose was 3.9 mmol/l (reference interval 3.5–5.3), plasma sodium 129 mmol/l (135–147), potassium 5.2 mmol/l (3.5–5.0), urea 17.8 mmol/l (3.3–6.6), creatinine 36 mmol/l (30–74), bicarbonate 15 mmol/l (21–28), ALT 15 mmol/l (<4 to +2) and C reactive protein 4 mg/l (0–5). Liver function tests and clotting were normal. She was resuscitated with a total of 30 ml/kg of colloid and crystalloid. The following day she relapsed with abdominal distension, shock, and deteriorating conscious level. Investigations showed glucose 14.2 mmol/l, amylase 20 IU/l (8–85), AST 186 IU/l (10–45), and ALT 129 IU/l (10–40). An x ray examination of the abdomen showed free air under the right hemidiaphragm. Emergency laparotomy revealed a single, 1 cm x 1 cm acute perforation in the second part of the duodenum. Histology and rapid urease test (CLIO) of the duodenal biopsy for Helicobacter pylori were negative. Fasting blood gastrin was 20 nl/l (10–100). She was discharged home taking omeprazole. Upper gastrointestinal endoscopic biopsy (eight weeks later) for histopathology and CLO test from oesophagus, stomach, antrum, and duodenum were normal.

Analysis of urinary organic acids by gas chromatography and mass spectrometry, obtained a day after clinical presentation, revealed a marked increase in 5-hydroxoyhexanoic acid (21% of total organic acids); a modest dicarboxylicaciduria (suberic accounted for 8% and adipic 6% of total organic acids); and a small but significant quantity of hexanoyl glycine (2% total organic acids) in the absence of ketonuria.

Blood obtained a week after a clinical presentation, when analysed by tandem mass spectrometry, showed octanoylcarnitine 0.19 mmol/l (<0.19), hexanoylcarnitine 0.67 mmol/l (<0.29), and decanoylcarnitine 0.63 mmol/l (<0.10), with a subnormal concentration of acetylcarnitine 4.0 mol/l (6.2–27.5). This profile was consistent with MCADD. Polymerase chain reaction/restriction digests based method revealed two mutations in the MCAD gene.

The clinical detail coupled with the absence of ketones and the increased 5-hydroxoyhexanoic acid led us to look for an abnormality in the oxidation of fatty acids, and resulted in identification of the minor constituent, hexanoylglycine that is recognised as an indicative marker of MCADD. Increases in urinary hexanoylglycine and 5-hydroxoyhexanoic acids in the absence of ketonuria have been reported previously in MCADD patients during clinical attack, and also in a boy who died. Our case was unusual in that the amount of 5-hydroxoyhexanoic acid was greater than even the sum of the individual dicarboxylic acids present, although high levels of 5-hydroxoyhexanoic acids are reported in acute episodes. The increased concentration of octanoyl carnitine in blood was also consistent with a diagnosis of MCADD.

We believe that this is the first report of MCADD presenting with duodenal ulcer. It could be argued that the ulcer was the primary problem and that the decompenasion was caused by the subsequent illness.
Thus, any child who has unexplained encephalopathy, regardless of its cause and clinical setting, should be screened for MCADD.

V Kairamkonda, M Dalzell
Department of Gastroenterology, Royal Liverpool Children’s NHS Trust, Alder Hey Children’s Hospital, Liverpool, UK

P D Losty
Department of Surgery, Royal Liverpool Children’s NHS Trust, Alder Hey Children’s Hospital

C Davidson
Department of Metabolic Medicine, Royal Liverpool Children’s NHS Trust, Alder Hey Children’s Hospital

Correspondence to: Dr M Dalzell, Department of Gastroenterology, Royal Liverpool Children’s NHS Trust, Alder Hey Children’s Hospital, Liverpool L12 2AF, UK; mark.dalzell@hct.nwest.nhs.uk

References

Glucose metabolism in sleep disordered breathing

An association between sleep disordered breathing (SDB) and impaired glucose tolerance has been reported in adults.1 Although SDB has been reported in diabetic children, no data are available on glucose metabolism in children with SDB. We used glycated haemoglobin (HbA1c) for the preliminary assessment of glucose metabolism in paediatric SDB patients.

HbA1c was measured in 12 children aged 26–116 months (mean 63) with suspected SDB owing to adenotonsillar hypertrophy. Informed consent was obtained from the guardians of each patient, and consent was obtained from the child if older than 5 years of age. Overnight polysomnographic studies were performed once for each patient by the standard method described elsewhere.2 The desaturation time (percentage of total sleep time with oxygen saturation <90%) minimum oxygen saturation level, and apnoea-hypopnoea index (AHI) were calculated. Complete blood count, blood gases, and blood chemistry (glucose, total protein, albumin, urca nitrogen, creatinine, uric acid, sodium, chloride, potassium, calcium, phosphates, lactate dehydrogenase, glutamic oxaloacetic transaminase, glutamic pyruvic transaminase, γ-glutamyl transpeptidase, alkaline phosphates, total bilirubin, total cholesterol, and triglycerides) were also determined.

The patients had no respiratory failure, heart failure, or coma. None of their weights exceeded 120% of their ideal weight for their heights. Desaturation time clearly divided the patients into two groups: six patients whose desaturation time was 0 or 0.1 (mild SDB group); and six whose desaturation time exceeded 4.0 (severe SDB group). The average HbA1c value for the severe SDB group (5.0, SE 0.07) was significantly higher than that for the mild SDB group (4.6, SE 0.10) (p = 0.01), although the actual HbA1c values were all within normal range. No other items showed significant differences between the two groups.

The severity of respiratory disturbances during sleep in diabetic children has been known to correlate with the duration of diabetes and with the HbA1c value.3 Recently, SDB parameters were found to be associated with worsening insulin resistance independent of obesity in adults.4 The current study shows that serum HbA1c is increased in association with the degree of desaturation in non-obese paediatric SDB patients; HbA1c levels should, however, be monitored after treatment. SDB and glucose metabolism are hypothesised to be closely associated in children as well as adults.

J Kohyama, T Hasegawa, J S Ohinata
Department of Pediatrics, Faculty of Medicine, Tokyo Medical and Dental University, Japan

Correspondence to: Dr J Kohyama, Department of Pediatrics, Faculty of Medicine, Tokyo Medical and Dental University, 1-5-45 Yushima, Tokyo 113-8519, Japan; jkohyama.ped@tmd.ac.jp

References

Short versus standard duration antibiotic treatment for UTIs: a comparison of two meta-analyses

Having recently published a meta-analysis on the same clinical question,1 it was with great interest that we read Michael et al’s systematic review of short versus standard duration antibiotic treatment for urinary tract infections (UTIs) in children.2 Given the publication (in close succession) of two meta-analyses on the same question with (on the surface) strikingly different explanations and conclude that the two divergent results was the different ways of treatment failure could have failed to capture the therapeutic advantage of standard duration treatment.3 Thus, our meta-analysis demonstrates that single dose or single day antibiotic treatment is not as effective as long-course treatment with shorter-course treatment even in recipients of short course treatments.4 The reasons for this exclusion are unclear, although we presume that they felt single-dose or single-day treatment was not a fair comparison with 7–14 day treatment. However, a number of randomised controlled trials (RCTs) made this comparison, suggesting that clinicians are, in fact, interested in the potential efficacy (and safety) of shorter-course therapies. Thus, our analysis is important, and our results add further weight to the evidence that short-course treatment is inferior to standard (7–14 days) treatment.5–7

The meta-analyses demonstrate that the divergent results were the result of how the treatment success was defined in the studies included. In our meta-analysis, we compared 3 days of treatment to 7–14 days of treatment, whereas Michael et al.2 compared 2–4 days of treatment to 7–14 days of treatment and excluded 11 studies comparing single-dose or single-day treatment to standard duration treatment.2,8,9

The reasons for this exclusion are unclear, although we presume that they felt single-dose or single-day treatment was not a fair comparison with 7–14 day treatment. However, a number of randomised controlled trials (RCTs) made this comparison, suggesting that clinicians are, in fact, interested in the potential efficacy (and safety) of shorter-course therapies. Thus, our analysis is important, and our results add further weight to the evidence that short-course treatment is inferior to standard (7–14 days) treatment.5–7

The meta-analyses demonstrate that single-dose or single-day antibiotic treatment is not as effective as long-course treatment for UTIs in children. The two meta-analyses together suggest that (1) "longer" short-course therapies may be as effective as 7–14 days of antibiotics and
(2) there is probably a duration of treatment threshold for “short-course” antibiotic treatment, above which longer duration of treatment confers no therapeutic advantage. Michael and colleagues suggest that as little as 2 days of treatment may be sufficient. However, only one of the trials in their meta-analysis studied 2-day treatment and that only ensured long-course treatment with a RR of UTI 0–7 days after completing short course treatment of 2.17 (95% CI 0.48 to 9.76). The duration of treatment threshold may be 3 days, but the point estimate of relative risk of treatment failure with 3 day treatment in our meta-analysis suggests otherwise. If the duration of short-course treatment for which there is no difference in efficacy compared with standard treatment is actually greater than 3 days, then the added convenience and cost-savings of “short-course” treatment become marginal. In the absence of appropriately powered RCTs (or meta-analyses) examining outcomes (treatment failure, reinfection, emergence of resistant organisms and cost) with “longer” short course treatment regimens (3, 4, and 5 days), we think that clinicians should continue to treat UTIs in children with at least 7 days of antibiotics.

R Keren
Department of Pediatrics, The Children’s Hospital of Philadelphia, USA

E Chan
Department of Pediatrics, The Children’s Hospital of Boston, USA

Correspondence to: R Keren; keren@email.chop.edu

References

Table 1  Results of three systematic reviews of randomised controlled trials comparing short duration with standard duration of antibiotic treatment for lower tract urinary tract infection.

<table>
<thead>
<tr>
<th>Systematic review</th>
<th>Comparison of duration of therapy</th>
<th>Number of data sets</th>
<th>Risk for persistent bacteriuria</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tran et al, 2001</td>
<td>1–4 days v &lt;5 days</td>
<td>13</td>
<td>RR 4.26 (95% CI 0.95, 9.48)</td>
</tr>
<tr>
<td>Keren &amp; Chan, 2002</td>
<td>3 days v 7–14 days</td>
<td>5</td>
<td>RR 1.36 (95% CI 0.68, 2.72)</td>
</tr>
<tr>
<td>Michael et al, 2002</td>
<td>2–4 days v 7–14 days</td>
<td>8</td>
<td>RR 1.06 (95% CI 0.64, 1.76)</td>
</tr>
</tbody>
</table>

*RR*, risk difference; *CI*, confidence intervals; *RD*, risk for persistent bacteriuria

Authors’ reply

In response to Keren and Chan’s thoughtful letter regarding our recent systematic review, we need to emphasise that the study question we addressed was different from that addressed by Keren and Chan in their own systematic review of randomised controlled trials comparing short with standard duration treatment in the treatment of children with urinary tract infection (UTI). The aim of our study was to determine the relative efficacies of short (2–4 days) and standard duration (7–14 days) treatment with the hypothesis that short duration may be as effective as standard duration treatment and provide potential advantages such as improved compliance. Therefore, we did not include trials in which single dose treatment was compared with standard duration treatment. In addition we chose to limit the review to trials in which the same antibiotic was used to treat each group, to avoid confounding.

The response to single dose treatment appears different from short course, suggesting that it is inappropriate to pool studies comparing single dose and standard treatment with those comparing short course and standard treatment. Three systematic reviews have now demonstrated that there is no significant difference in the number of children with persistent bacteriuria after short duration or standard duration treatment (see table 1). In contrast, Keren and Chan found that significantly more children had persistent bacteriuria following single dose compared with standard duration treatment (7 data sets: RR 2.73, 95 CI 1.38 to 5.40). Similarly, Tran et al in their meta-analysis of 22 studies comparing both single dose and short duration treatment with standard duration treatment found the latter to be more effective (risk difference 6.38; 95% CI 1.88 to 10.89).

Because there is no significant difference between short duration and standard duration treatment in the number of children with persistent UTI after treatment, it is not possible to calculate a number needed to treat to prevent one episode of persistent bacteriuria.

From our systematic review, we are not able to determine whether there is an “optimum duration of treatment threshold” as postulated by Keren and Chan. Only one study included in the meta-analysis, examining the effects of short duration and standard duration treatment in clearing bacteriuria, compared 2 days of treatment with 10 days’ treatment. In their letter above, Keren and Chan argue that this study favours standard duration treatment. However, there was no significant difference between treatments in the number of children with persistent bacteriuria at the end of treatment (RR 2.17; 95% CI 0.48 to 9.76) although the wide confidence intervals do not exclude the possibility that short duration treatment could be more or less effective than standard duration treatment.

No significant differences in the number of children with persistent UTI after treatment between short duration and standard duration antibiotic treatment have been found in three systematic reviews of randomised controlled trials despite different study inclusion criteria and definitions of persistent infection. As described in our review, the wide confidence intervals around the summary estimates indicate residual imprecision in the results. However, this statistical imprecision is of doubtful significance for most children, who are at a low risk (1–3%) of persistent UTI at the end of treatment following their first lower tract UTI. Therefore, we do not support Keren and Chan’s conclusion that clinicians should continue to treat lower tract UTI with standard duration treatment. Instead, we believe that short duration treatment may be used to treat children with lower tract UTI.

E M Hodson, M Michael, J C Craig, S Martin
Centre for Kidney Research, The Children’s Hospital at Westmead, Sydney, Australia

V A Moyer
Center for Clinical Research and Evidence Based Medicine, The University of Texas–Houston Health Science Center, Houston, TX, USA

Correspondence to: E Hodson; Elisas@chw.edu.au

References

www.archdischild.com
Is life long follow up for patients with Kawasaki disease indicated?

Brogan et al recommended life long follow up for patients with Kawasaki disease, including those who have not had coronary artery involvement. The reason they quoted was to document the blood pressure and provide general advice regarding other risk factors.3 The American Heart Association recommends echocardiographic (Echoc) evaluation of the coronary arteries at presentation and follow up ECG at 6–8 weeks and 6–12 months after the onset of symptoms for those who did not have or just have transient coronary artery involvement. They do not recommend follow up after first year unless cardiac disease is suspected.4

Tuothy et al demonstrated, in their multi-institutional review of 536 patients, that no patient with a normal follow up ECG, performed within 2 months following disease onset, subsequently developed echocardiographic coronary artery abnormalities. Even those patients with initial echocardiographic abnormalities that became normal at 1–2 months remained normal thereafter.5 Scott and colleagues showed that no patient with a normal ECG at 2 weeks to 2 months after the onset of symptoms had subsequent ECGs that revealed coronary artery abnormalities, and questioned the value of 6–12 month ECG in the same group.6

Brogan et al did not make any comments about the adverse effects of life long follow up such as anxiety and inappropriate restriction of activities. Finally, there were no comments about the cost and resources for providing life long follow up. The authors did not specify whether paediatric cardiologists, general paediatricians, or general practitioners would follow up; all of them already have increasing demands of workload.

S J Murugan, J Thomson, J M Parsons
Yorkshire Heart Centre, Leeds General Infirmary, Leeds, UK

Correspondence to S J Murugan; jothdevi1@hotmail.com

References

Management of childhood osteoporosis

I read with interest this recent review article that summarises current knowledge about this subject. I have a number of comments that are pertinent to the discussion. As the authors allude to, there is currently a lack of good evidence on which we can base preventive management. Although calcium and vitamin D supplements are routinely used by some paediatric rheumatologists, there appears to be only one short term study suggesting this may be beneficial for bone density.7 There is a lack of evidence in relation to growth hormone therapy are methodologically flawed because neither have accounted for the change in apparent bone density, which will occur in any child who grows better for any reason when assessed by modalities such as dual energy x ray absorptiometry.8

As illustrated by another article in the August 2002 edition of Archives, there is a lack of good evidence on which to base much paediatric management and it is imperative that further research, especially randomised controlled trials, is undertaken in the area of prophylaxis against osteoporosis in children with chronic disease on steroids. Paediatric endocrinologists will be familiar with the flurry of small uncontrolled studies undertaken in numerous groups of children with chronic disease in which calcium and vitamin D supplements are given. Many reports of short term improvements in growth velocity have not been supported by long term outcomes in height. There is a risk that a similar phenomenon will occur with the use of bisphosphonates in children with chronic disease and low bone density without properly designed studies and satisfactory outcome measures.

The use of glucocorticoids in children with chronic disease occurs across many paediatric subspecialties and I would argue strongly that the management and prevention of osteoporosis requires specialist expertise just as the management of growth retardation currently does. It is important that in each tertiary centre such a specialist service is provided by one department that has expertise in the interpretation of bone density scans in children and the management of children with osteoporosis. Such individuals may not only be paediatric endocrinologists but may be a paediatric rheumatologist, a general paediatrician with a special interest in bone disease or a metabolic bone disease subspecialist. It is only in this way that we can learn more about the management of this condition and avoid children being treated inappropriately.

N J Shaw
Birmingham Children’s Hospital, Birmingham, UK
nick.shaw@childrens.wmids.nhs.uk

References

Newborn screening for Duchenne muscular dystrophy

Elliman, Dezateux, and Bedford,1 in their recent leading article on newborn and childhood screening, include reference to newborn screening for Duchenne muscular dystrophy (DMD). They argue that the main value of such a screening programme is to warn parents that future sons may be affected, and support this statement with reference to Jarvinen et al.2 This paper does not report a newborn screening study but the results of a retrospective study of 23 females in Finland carrier tested for DMD during childhood. However, a newborn screening programme for DMD has been running in Wales since 1990 (1990–8 as a research evaluation and from 1998 health authority funded). During the research period interim evidence was published.3 More recently the full results of our prospective study have been published.4 Our evaluation has demonstrated that a newborn screening programme for DMD can be acceptable to both parents and health professionals, providing that a rigorous service delivery protocol is in place and the programme is supported by an effective infrastructure, in particular by paediatric and genetic services.

E P Parsons
SONAMS and Institute of Medical Genetics, University of Wales College of Medicine, Cardiff, UK

D M Bradley
Department of Medical Biochemistry, University Hospital of Wales, Cardiff, UK

A J Clarke
Institute of Medical Genetics, University of Wales College of Medicine
Correspondence to Dr Parsons; parsonsep@cf.ac.uk

References

www.archdischild.com
The effect of sanctions on children of Iraq

Sanctions were imposed on the people of Iraq in 1990. Iraqi people are still suffering, especially children. Infant mortality (IM) has increased more than five times. Previously it had decreased from 139 in 1960 to 20 in 1989, which was comparable to developed countries. In 1992 it went up to 111.1 In 1999, a decade later, IM was still high at 104.2 The Gulf War and trade sanctions caused a three-fold increase in mortality among Iraqi children under 5 years of age. It has been estimated that more than 46,900 children died between January and August 1991.3

The study of the UN Food and Agricultural Organisation, published in a letter to the BMJ in 1995, concluded that deaths of more than 50,000 children could be attributed to UN sanctions. It also stated that the death rate among children under 5 years in Baghdad had increased fivefold since the war ended in 1991.4 Data for 1994–99 showed that mortality under 5 years was 131 per 1000 live births, compared with 56 for 1984–9, and the mortality for children under 5 years was 131 per thousand. A WHO investigation in 1995 suggested a possible link to products—now banned—from familiar markets, which were derived from depleted uranium used in piercing artillery shells. There were staggering deficiencies in cancer treatment facilities because of UN sanctions which were intended to exclude food and medicines.

A report in 1996 showed that one third of hospital beds were closed. More than half of all diagnostic and therapeutic equipment was not working due to lack of spare parts and maintenance. All public hospitals experienced serious problems with lighting, cleaning, water supply, and sewage. The population had been burdened by a rapid rise in serious infections, nutritional deficiencies among children and pregnant women, and other treatable conditions for which neither drugs nor operations were available.5

Paediatricians have been isolated by the intellectual embargo from the international medical community. Physicians who wish to attend international conferences face travel restrictions, like denial of visas to European countries or the USA. In 1990, the delivery of European and American journals were abruptly stopped. This intellectual embargo served to undermine the care of patients, and denies Iraqi doctors the right to share scientific advancement and its benefits.6

References


hypoxic vasoconstriction. Therefore Dr Casano’s recommendation for the early use of pulmonary vasodilators is unlikely to be sufficient in this context. We are assessing the impact of strategies aimed at reducing lymphocyte numbers and adhesion in addition to standard treatments for pulmonary hypertension.

M J Peters, C M Pierce  
Paediatric Intensive Care Unit, Great Ormond Street Hospital, London, UK

N J Klein  
Infectious Diseases and Microbiology Unit, Institute of Child Health, London, UK

Correspondence to: Dr Peters; m.peters@ich.ucl.ac.uk

References

Authors’ reply
As Peters comments in his letter, we know that hyperleukocytosis has been postulated as a factor for pulmonary hypertension in Pertussis infection, but necessary brevity did not make it possible to report. Nevertheless, our patient never reached these values of leukocytosis; it’s possible, as in many other diseases, that several pathogenic mechanisms contribute to pulmonary hypertension, making a concomitant treatment approach necessary.

M Pons, P Casano  
Hospital Sant Joan de Déu, Unidad de Cuidados Intensivos Pediátricos, Passeig de Sant Joan de Déu, 2 080950, Esplugues de Llobregat, Barcelona, Spain

Correspondence to: Dr Pons; mpons@hsjdbcn.org

CORRECTIONS
In the paper by Clarkson and Choonara in the December issue of ADC (Arch Dis Child 2002;87:462–7) the following corrections have been noted:

Results; first sentence: there were 331 deaths with 390 suspected drugs (not 390 and 389 respectively as stated in the paper).

Results; section “Corticosteroids”: the third sentence starting “No details were available...” should be deleted.

Results; section “Non-steroidal anti-inflammatory drugs (NSAIDs)”: the second sentence “All reports for NSAIDs have occurred since 1990” should be deleted.

Discussion; fifth paragraph: the penultimate sentence should be “as recently as 1999 our study found a single fatality” (not 2 reported fatalities).

Figure 1  Scimitar syndrome. Chest x ray showing a curvilinear density which extends from the right hilum towards the right hemi-diaphragm which represents the anomalous pulmonary vein.

The journal apologises for the errors.