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Clinical presentations

An Evolving Pituitary Endocrinopathy

A Duncan, P J Smail, G F Cole

A 12 year old girl presented with worsening of primary enuresis associated with a large fluid intake, obesity, tall stature and a family history of sarcoid. Initial investigations confirmed cranial diabetes insipidus and hypothyroidism. Neuro-imaging was normal.

She was treated with DDAVP and thyroxine and observed carefully. She developed a blistering, ulcerative vulval rash which proved resistant to treatment and was biopsied showing only chronic inflammatory changes. Despite further neuro-imaging, not until 18 months later was a lesion demonstrated in her pituitary on CT and MRI scans. A stereotactic biopsy was done with some difficulty and suggested a probable diagnosis of Langerhans' cell histiocytosis. There was no evidence of involvement of other tissues.

Her anterior pituitary function had deteriorated over this time and there was now evidence of gonadotrophin and adrenal insufficiency. Despite treatment with growth hormone and full gonadal replacement in addition to hydrocortisone, her weight has increased to over 160 kg.

A small left visual field defect has now developed and this has prompted treatment with radiotherapy to the pituitary region. Even if local disease control is achieved and hormonal replacement is optimised, the patient may be left with morbid obesity as a long term threat to her health.

This case demonstrates that the onset of diabetes insipidus in childhood, followed by an evolving anterior pituitary endocrinopathy, requires careful monitoring until an occult intracranial lesion declares itself.

Citrobacter Meningitis in Infancy

C A Stuart, M Morgan, G F Cole

Two cases of citrobacter diversus meningitis are described. Citrobacter is a widespread gram negative bacillus and citrobacter diversus has been recognised as a cause of meningitis in babies since the 1970s. The illness is especially devastating because of the high incidence of cerebral abscess formation.

The first baby presented at 10 days with a septicaemia. Citrobacter diversus was isolated from the CSF. She was treated with intravenous Cefotaxime and Gentamicin for 6 weeks. At 24 hours her cerebral ultrasound appeared normal. At 7 days she had widespread patchy oedema and a large right frontal abscess cavity. After a further week a second abscess had appeared in the left frontal lobe. Both abscesses required to be tapped to relieve raised intracranial pressure on 4 occasions. A ventriculo-peritoneal shunt was inserted at 6 weeks because of increasing hydrocephalus.

The second infant presented at 5 weeks with evidence of raised intracranial pressure. On CT scan she was found to have a left frontal abscess. Citrobacter diversus was isolated from the abscess fluid. She too was treated with a prolonged course of intravenous antibiotics. The abscess required regular tapping over the first 2 weeks. Hydrocephalus requiring shunting developed later in the illness.

Subsequently both babies have had delayed development - not surprisingly a common sequel to citrobacter diversus meningitis.

A CASE OF NEUROFIBROMATOSIS 1(NF1)-NOONAN PHENOTYPE AND THE IMPLICATIONS FOR GENETIC COUNSELLING IN CASES FOR ADOPTION

P.D. Turnpenny[1], J.C.S. Dean[1], D.J. Lloyd[2] and G. Russell[2]

A 29 year old woman with dysmorphic features, short stature, widespread neurofibromata and previous surgery for pulmonary stenosis, gave birth to a female infant who at seven months had five cafe-au-lait patches. A diagnosis of NF1-Nanoon phenotype in the mother was made in the post partum period. Eight years previously she had given birth to a female infant with mild dysmorphic features and a heart murmur, which was adopted. At that time the possibility of maternal NF1 was raised but not confirmed and was not included in the adoption medical report. The adopted child is reported to have had corrective surgery for ptosis, suggesting she has the Noonan phenotype, but no other details are available. It is unlikely that she has been examined specifically for either pulmonary stenosis or cafe-au-lait spots.

In the light of a newly diagnosed genetic condition, medico-legal and ethical dilemmas, as well as issues of confidentiality, are attached to the question of counselling the adoptive parents and later the adopted child when she reaches reproductive age. There are implications for a re-evaluation of the role of family history, examination and genetic counselling in adoption cases.

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A Bizarre Encephalopathy?

E J Ashcroft, G F Cole

CASE HISTORY A previously well, fully immunised 8 year old boy developed a bizarre picture of neurological signs and symptoms over a 5 week period. There were focal and generalised fits and left upper motor neurone signs progressed to bilateral signs. Bulbar palsy and aphasia developed. Dystonic movements occurred and increased until he had frequent oculo-gyric crises and opisthotonic spasms. There were severe, frightening visual hallucinations. Symptom control was finally achieved with intravenous anaesthesia (Propofol). The prevailing view was that this was a rapid neurodegenerative process with an appalling prognosis. However slow improvement began in week 7 and the Propofol infusion was withdrawn gradually. Upon discharge home in week 13, he was weak, incoordinate, had severe short term memory loss and immature behaviour. The boy is now neurologically normal and has no apparent learning difficulties.

INVESTIGATIONS Viral studies on CSF and serum were negative. Mycoplasma, Borrelia Sp., Brucella, Rabies, Tetanus and TB were considered. A drug screen was negative and serum heavy metal levels were normal. As there were several neurotoxic compounds in the household, Munchausen's by proxy was considered: this suspicion was never entirely resolved. Neurodegenerative disorders were excluded on neuro-imaging, biochemical assays and normal brain biopsy. Normal bone marrow, urine screen and peripheral blood film excluded storage disorders. Autoantibody studies were negative. Organic and amino acids were normal and a porphyria screen was negative. Serum lactate and pyruvate were normal. EMG studies were normal. EEGs showed right sided asymmetry only.

In summary this 8 year old child had an unexplained life threatening neurological disease. His recovery appears to be complete and he has no recall of his illness.

Scientific presentations

EARLY PREDICTION OF CHILD ABUSE: WHAT CAN THE MATERNITY HOSPITAL CONTRIBUTE?
M.S. Fraser, V.J. Linnemann

Ninety children, abused before the age of 3 years were identified and their birth records studied. 45 had suffered physical abuse and 45 serious neglect; two controls for each were added. The aim was to derive, from details routinely found in obstetric records, a simple score by which family doctor and health visitor could be alerted to families with greater than average need for support and guidance.

Analysis by stepwise logistical regression selected 7 factors which with simple weighing would produce a score ranging from 0 to 12. A score of 3 or more identified 64.5 per cent of the abused group, with 6 per cent of false positive scores among controls. This could serve as a risk marker, needing a delicate approach to the families.

The factors generated were: mother under 20 years; smoking in pregnancy; partner unemployed; his occupation not recorded; few antenatal visits; mother discharged herself from hospital against advice; mother of low intelligence. By this method the 90 abusing families would score 0 to 10, mean 3.52, and 180 controls 0 to 5, mean 0.82.

There were few differences between the perinatal antecedents of physical abuse and those of neglect. In this study the dozen highly significant differences, found between abused and control groups, did not include the circumstances commonly thought to interfere with mother-infant bonding.

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NONINVASIVE COMPARISON OF RESULTS OF PULMONARY VALVOTOMY AND TRANSCUTANEOUS BALLOON VALVOPLASTY
J.F.M. Bruinenberg, A.B. Houston, J. Burns, J.W. Kay

Transcutaneous balloon valvoplasty is the recommended procedure for isolated pulmonary valve stenosis. We used imaging, spectral and colour Doppler ultrasound to compare the results of transcutaneous balloon valvoplasty (30 patients) and surgical pulmonary valvotomy (28 patients) with particular reference to the residual transvalvar gradient and the degree of pulmonary regurgitation.

Gradient distribution before intervention was statistically comparable in both groups. There was a statistically greater reduction in the transvalvar gradient in the surgical group (from 65.2 + 37.9 to 13.1 + 6.5mmHg after a follow up of 2.0 + 0.9 years). However the surgical group showed a greater degree of pulmonary regurgitation as assessed by diastolic backflow in the pulmonary artery and larger right ventricular size. These results justify the continued use of transcutaneous balloon valvoplasty as treatment of isolated pulmonary valve stenosis. The lesser degree of residual regurgitation may be an advantage in the long term.

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COMPUTER ANALYSIS OF PHYSIOLOGICAL DATA IN A NEONATAL COMPUTERISED COT MONITORING SYSTEM
S. Cunningham S. Deere CA. Bass N. McIntosh

Computers monitoring physiological parameters play an increasing role in intensive care areas. Progress will involve developing computer intelligence. We are combining a series of data analysis techniques to identify two main types of event in a multichannel physiological computerised cot monitoring system. (1) Events due to intervention with equipment or poor quality signals i.e.artifact. Within these, predictable events are the majority. (2) Events due to a pathological change in the baby's physiological status. We apply the same principles to both types of event. Central to the method is the development of a 'model' of the event. This is achieved by manipulation of data using the following techniques. (a) Exclude values outside a specific range. (b) Note unacceptable rates of change; Single channel or Combination of channels. (c) Smooth data using a BOX filter. (d) Take into account the temporal relationships of associated parameters in an event. A derived model is event specific. Once the model is identified, the computer can exclude those values from further analysis if due to artifact, e.g. a pO2 probe change, or can give early warning of a pathological event e.g.an ET tube blockage.

We are able to conclude four main points from our work so far; (1) Data from real time neonatal monitoring systems are too variable to exclude all artifact. (2) The majority of artifact can be removed by relatively simple measures. (3) Complete removal of all artifact has an exponential increase in difficulty. (4) Computer identification of pathological events is possible using the same methods as those for artifact rejection.

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A NEW GENERATION COMPUTER MONITORING SYSTEM
McIntosh N, Bass CA, Badger PG

The extra dimension of a time base for physiological monitoring provides additional information which allows diagnosis and treatment to be adjusted more critically. Such trend monitoring is at most only a minor component of current monitors and charting on paper is used to build up an intermittent picture of the developing pathophysiology.

MARY3 is a computer controlled monitoring system which automatically samples and records data from intensive care patients. Up to 32 channels of physiological and environmental data can be sampled every second from primary patient monitors, ventilators and other sources. The sampled values are averaged each minute and recorded by the system on a hard disk. The system can record the 1-second data continuously or as a retrospective snapshot of an event in patient care. Short periods of waveform data can be sampled, viewed and recorded. Medical and nursing comments can be added using the computer keyboard. The system provides screen layouts for the manual input of other data e.g. blood gas, ventilator settings, fluids and drugs.

The sampled data can be plotted as trend graphs on the computer screen in realtime and all the data recorded from the beginning of a patient stay can be recalled for display and printing. Graph sizes and scales can be changed and trend data can be displayed over timescales ranging from 6 minutes to 3 days. Artifact data can be removed and hourly mean, standard deviation and other data analyses can be displayed. There is a database facility which keeps a detailed record of the patient stay for statistical and audit purposes.

At Edinburgh the 12 NNICU cots are networked to this computerised system with the local recorded data also routed for display at a central doctors station.

MARY3 is valuable for day to day clinical care with the added benefit that all the recorded data is available for research. In the future MARY systems will be able to recognise changing patterns in the sampled data and give an early warning of a deteriorating situation.