Acrodynia: a case report of two siblings

Acrodynia, a rare disorder, is a form of chronic mercury poisoning.1 We report two siblings who developed the classic clinical picture of acrodynia.

A 4½ year old boy was admitted with dysuria, general weakness, and loss of appetite. He had hypertension (140/95 mm Hg) and tachycardia (141 beats/min). He was irritable and depressed, and had a diffuse itching papular rash with palm erythema and superficial desquamation (fig 1). Initial evaluation revealed a normal complete blood count and a normal blood chemistry. Urine analysis and complement levels were normal. Vanillylmandelic acid in a 24 hour urine collection was 22.2 µmol/day. Duplex scan of the renal arteries, abdominal ultrasound, and computed tomography (CT) of the chest and abdomen, were all normal. Heart echocardiography showed mild hypertrophy of the myocardium. TSH was 5.53 mIU/L, and free thyroxine 24.45 pmol/L. A brain CT scan revealed a point calcification at the right caudate nucleus and several bilateral areas of low density in the white matter. EEG was normal. A successive complete blood count revealed haemocytoma (haemoglobin 165 g/l and haematocrit 48.1%).

After eight days, the patient’s 6 year old brother was admitted with general weakness, pain in his lower extremities, and a diffuse itching papular rash with palm erythema and superficial desquamation. He was hypertensive (126/87 mm Hg) and tachycardiac (140 beats/min).

Due to the fact that both siblings presented, at the same time, with more or less the same complaints and physical findings, it was suspected that their condition may have been the result of an environmental exposure. It was discovered that three months previously, the children had played with a broken sphygmomanometer for a few weeks.

Urine mercury level for patient 1 was 158 µg/g creatinine and for patient 2, 113 µg/g creatinine. Urine mercury level for patient 1, after a dose of captopril (chelating agent), was 214 µg/g creatinine. Chelation was initiated with dimercaptoprosonic acid for a 19 day course. Two weeks later, symptoms had almost resolved and the rash disappeared. A month later, blood pressure and heart rate had returned to normal.

Torres and colleagues published a review of eight cases of acrodynia. In all these cases, and in ours, the physicians first thought of phaeochromocytoma. Mercury inactivates an enzyme that participates in the breakdown of catecholamines, and therefore their concentrations increase, stimulating a phaeochromocytoma like syndrome.

Torres and colleagues also reported that in two of the patients reviewed, haemocoencentration was observed, most probably due to intravascular and extracellular volume depletion. This was also found in our patients.

The brain CT findings of low density in the white matter, in patient 1, were not specific. Neurological examination was normal apart from mental changes that are common in acrodynia. To the best of our knowledge, this is the first time that abnormalities in brain CT have been described in acrodynia.

In summary, acrodynia, although rare, should be considered in every child presenting with hypertension, tachycardia, mental changes, and cutaneous manifestations. This case emphasises the fact that good history taking is an essential element in even the most puzzling clinical pictures.

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References

Bordetella pertussis infection causing pulmonary hypertension

In Spain the incidence of whooping cough is less than five cases per 100 000 inhabitants.1 Mortality rate is 0.4% in the United States, heart failure being one of the most frequent causes.2 Although cases of death due to Bordetella pertussis infection as a consequence of lung hypertension have been described previously,3 this complication is not very well known. Here we report a recent case and review the literature.

A 23 day old girl, who had had a pertussis cough for several days, was admitted with breathing difficulty of 12 hours duration. On admission she had tachycardia (heart rate 180 beats/min), tachypnoea (70 breaths/min), pyrexia (38°C), and haemoglobin saturation 90% without oxygen. A chest x ray revealed right superior and half lobe infiltrates. Blood analysis showed 33 x 10⁷ leucocytes with left deviation and 33 mg/dl C reactive protein. Testing for respiratory syncitial virus was negative, direct immunofluorescence and culture for B pertussis were both positive.

The patient was admitted and treated with oxygen and erythromycin. After 12 hours she developed respiratory failure (respiratory rate 100 breaths/min, pH 7.16, pco₂ 74 mm Hg, po₂ 57 mm Hg, HCO₃⁻ 26, base excess –5) and was transferred to the paediatric intensive care unit with intubation and pressure control (peak inspiratory pressure 22, peak end expiratory pressure 3, Fio₂ 0.35, respiratory rate 40). Twenty four hours later, hypoxaemia necessitated increasing Fio₂ to 1, and refractory hypotension required volume load and inotropics (TAM 38). Echocardiography diagnosed severe lung hypertension (pulmonary artery pressure 65 mm Hg) and decreased heart contractibility. Nitric oxide 8 ppm and milrinone 0.37 µg/kg/min temporarily improved po₂, but this subsequently deteriorated (po₂ 40 mm Hg). High frequency ventilation therapy was initiated; nitric oxide up to 20 ppm was given, and inotropic support enhanced, but with no response. She suffered a fatal cardiac arrest 98 hours later. The family did not authorise necropsy.

Fatal myocardial failure secondary to lung hypertension has been reported in four infants under 2 months with verified B pertussis infection.4 All presented with initial tachycardia (160–220 beats/min) refractory to treatment with volume load, and developed posterior persistent hypertension that did not respond to inotropic support.

Lung vasodilators such as nitric oxide, milrinone, or prostacycline may be useful in management of these patients, although they did not prove beneficial in our patient.

Because of the rapid deterioration of all these cases, we recommend early echocardiography diagnosis, enabling vasodilator
therapy to be initiated in the early phase of lung hypertension in order to improve prognosis.

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1 National Epidemiology Center: Immunization against whooping cough in Europe: situation at the end of 1999. Epidemiologic Bulletin of Spain, National Epidemiology Centre.

Sudden infant death syndrome: problems, progress and possibilities

As an internationally recognised disease classification, sudden infant death syndrome (SIDS) is unique in that the diagnosis is reached by exclusion, by failing to demonstrate an adequate cause of death. By definition it is imprecise, the diagnosis of SIDS depends on the thoroughness of the post mortem examination, the extent of detail given in the clinical history and the meticulous nature with which the death scene investigation is carried out. Even if these conditions are satisfied to some chosen specification, this is not an endpoint but a rather a beginning, as we are still left with the question of “why did these babies die?”

The tragedy of SIDS is not a modern phenomenon but was only christened a syndrome 40 years ago and, after extensive research, the possibility of finding a collection of symptoms and signs manifesting as a single cause appears extremely unlikely. Some experts suggest a triple risk causal mechanism for SIDS involving a vulnerable infant, a critical development period and an exogenous event that would not normally put a healthy child at risk. The controversy lies in what they see as a definition of consequence, in order to restrict the liberal use of such a diagnosis to exclude suspected cases of accidental suffocation and infanticide. Hypotheses continue to proliferate and, as the evidence for risk factors mount, the debate has widened from causation to the relative safety of accepted infant care practices.

In trying to understand how infants die, we have come to the realisation that we must first understand how infants survive. SIDS research has developed from basic epidemiological and pathological findings at death to a wider investigation of infant sleep structure, care practices, physiology, and genetics. This multidisciplinary approach is elegantly illustrated in Byard and Krouss’s book. The eclectic choice of contributing experts gives a clear insight into current thinking and recent discoveries in different fields, while challenging the reader with a subtle consensus of disagreement. The book gives detailed background of each debate but is more than a reference manual for other researchers in the field. Given the rarity of SIDS, many medical professionals may not have had experience with the sudden death of a child but will have to deal with mothers concerned about child safety, while some parents are reticent to accept advice unless they know how this has been derived, this book is also for them.

If there appears to be a lack of co-ordination in the approach among different research groups, a slightly over zealous interpretation of findings by some experts and perhaps more clarity than clarity in the overall picture, then this book has given a true reflection of SIDS research as it currently stands. There is no ending to the story because infants still die suddenly and unexpectedly, but if SIDS research is to be ultimately judged by the number of young lives so far saved then the endeavours of those involved should be highly commended.

P S Blair


Coming back to the new edition of this book is like coming back to an old friend. Like many paediatricians, I have used the first edition as a valuable reference in child protection cases. The expertise and experience of all three authors are well recognised internationally and there is no doubt that this edition will continue to be a valuable aid to all clinicians working with children.

All aspects of abuse are covered and there are helpful summaries in each chapter. It is an easy book to read but also I find it easy to get information on individual issues in child protection. There is an interesting historical introduction: although I would have liked rather more before modern times.

The problem I find with this book is that it is not really evidence based in a modern sense. Papers are quoted with no real attempt to assess their quality. This is partially because there are so few quantitative studies in child protection but I think readers would have liked to have more descriptions on the quality of the methodology of the papers that are quoted. I would have liked the references tabulated in each area of abuse. There are also concerns regarding the section on epidemiology of abuse. The histogram that is used as an illustration does not give incidence rates nor is it population based.

I particularly studied areas in the book that I know cause diagnostic difficulty and where there is controversy. One of these is subdural haemorrhage. I was disappointed that the section was quite short: only four pages. I was also disappointed at the number of references, only 14, in what is the most common cause of serious physical harm in physical child abuse.
I find that neglect and emotional abuse are areas where it is difficult to put facts together for a clear diagnosis. The section on neglect has a helpful list of points to look for in the potentially neglected child and also ways of assessing the whole family. I found the section on emotional abuse less helpful. Child protection is a very difficult area for clinicians and many shy away from committing themselves to clear diagnoses. This new edition will help give more confidence in dealing with these difficult cases. It is a pity that at nearly £70 it will not be accessible to young doctors outside libraries. Perhaps fewer photographs and being in paperback would make it less expensive and more accessible.

J R Sibert

Mosby’s atlas and text of pediatrics and child health


I enjoyed reviewing this book aimed at students and doctors in training, and I also learned from it. I must add that it is a good source of information for doctors who are preparing for examinations.

The book gives useful information, is highly illustrated and the format with text boxes and lists itself for easy reading and reference (revision for examinations).

The photographs are well placed with the text and with excellent explanations, which accompany the photographs, x rays, and scans. The quality of the photographs are superb too, thus the clinical phenotypes, which the authors want to illustrate are clearly visible. I found the book easy to read and understand.

I am sure that this book will prove very useful and will fill the gap in the market, as it will attract those adult learners who learn visually. It lends itself for scan reading for revision.

I teach examination preparation courses and I will bring this book to the attention of candidates sitting the DCH and MRCPCH exams. I would think that the GP tutors who come across this book would find it helpful in their teaching too. Many of the illustrations and slides will enhance anyone’s teaching methods.

More books like this are needed in paediatrics and child health as the pictures and illustrations that the doctors see will enhance their learning (and retention) skills. With problem orientated teaching (and learning) that we now practise, this type of book and presentations would be a most welcome addition. The market is not saturated, and I hope it will never be.

S Lingam

CORRECTION

Unfortunately the authors for the items in the Archimedes articles for September and November 2001 were not correctly coded and do not show up using searches on ADC Online or Medline. The authors for these articles should be cited as follows:

September


November

