

Malnutrition, Liver Dysfunction, Subdural and Retinal Haemorrhages and Encephalopathy in Children Resulting from a Deficiency or Abnormality of Vitamins C, D and K

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Abstract *Subdural and retinal haemorrhages, encephalopathy, bruises and fractures in children have hitherto been attributed to shaken baby syndrome, non-accidental injury, abusive head trauma or inflicted brain injury. The purpose of this investigation is to explore an alternative explanation. Since vitamin K and other nutrients are necessary for the integrity of both the coagulation of blood and the mineralization of bone, and since a deficiency of this vitamin is known to cause haemorrhagic disease of the newborn and fractures in children with cystic fibrosis and biliary atresia, it seems appropriate to investigate the status of vitamin K and other essential nutrients in children alleged to have suffered non-accidental injuries. It was found that all three children reported here had a prolonged prothrombin time with evidence of liver dysfunction and malabsorption/malnutrition as shown by abnormality of the liver enzymes and a reduced level of serum albumin. It is concluded that subdural and retinal haemorrhages, encephalopathy, bruises and fractures in children which have hitherto been attributed to shaken baby syndrome, non-accidental injury, abusive head trauma or inflicted brain injury is due to a metabolic disorder resulting from a deficiency or abnormality of vitamin K and other essential nutrients such as vitamins C and D following malnutrition/malabsorption or liver dysfunction.*

Introduction

In the mid 20th century following the publication of a report by an American radiologist on fractures and haemorrhages in infants¹ a neurosurgeon in England suggested that the cause of the lesions was violent shaking of the infant by an adult² and this claim was echoed by the radiologist³ and given the name “shaken baby syndrome” (SBS).

When it was realized that skull fractures could not be explained by shaking, a professor of paediatrics in England suggested that

a violent impact of the skull against a hard object was the most probable cause of this type of fracture.⁴ The SBS became known as “the shaken-impact syndrome.”

Retinal haemorrhages seen in these children were claimed to be conclusive proof of abuse and ophthalmologists added “acceleration-deceleration of the head as it is violently rotated by the abuser as the cause of the retinal hemorrhages.”⁵

Throughout academia the current teaching is that it is the combined “triad” of sub-

dural and retinal haemorrhage with brain damage, as well as the characteristics of each of these components that allow a reconstruction of the mechanism of injury, and assessment of the degree of force employed. The application of rotational acceleration and deceleration forces to the infant's head causes the brain to rotate in the skull. Abrupt deceleration, it is claimed, allows continuing brain rotation until bridging veins are stretched and ruptured, causing a thin layer of subdural haemorrhage on the surface of the brain.⁶⁻¹⁷

Maguire et al¹⁷ claim that their review, the largest of its kind, offers for the first time, a valid "statistical probability of inflicted brain injury" (IBI) when certain key factors are present.

One of the "key factors" upon which they base their opinion, retinal haemorrhages, is known to be associated with raised intracranial pressure from any cause as in Terson's syndrome¹⁸ and following vitamin K deficiency.^{19,20}

Apnoea, also, is rated high in their list of statistical markers of IBI and is claimed to be a crucial distinguishing feature. Apnoea is a feature of the condition known as an apparent life threatening event (ALTE), which can be caused by prematurity, gastro-oesophageal reflux, cardiac arrhythmia, laryngomalacia, trachiomalacia, infection, metabolic disorders and seizure and has been reported by Ghosh et al as "Shaken baby syndrome masquerading as an apparent life threatening event."²¹ Bruises, rib, spinal and limb fractures not "satisfactorily explained" have been attributed to physical abuse by the perpetrator, but Clemetson²² has shown that the clinical findings "were compatible with and even suggestive of infantile scurvy or toxic histaminaemia" and "bruises of the thigh and even fractures of the femur have been recorded as arising from the gentle act of diapering a scorbutic infant."²³

The following cases demonstrate the clinical features of the metabolic disorder, which is invariably associated with some or all of the following biochemical markers of malnutrition and/or liver disease:

1. Haemoglobin: decreased
2. Prothrombin Time (PT): increased
3. Albumin: decreased
4. Total protein: decreased
5. Aspartate aminotransferase (AST or SGOT): increased
6. Alanine aminotransferase (ALT or SGPT): increased
7. Gamma-glutamyl transpeptidase (GTT): increased

Case #1

The mother was found to be anaemic and was prescribed iron therapy during her pregnancy. The infant was born by a normal vaginal delivery and had Apgar scores of 9 and 10. His birth weight was 8 lbs 15 oz. He had a congenital right hydrocele.

Laboratory investigations performed the day after birth showed:

1. White blood count: $31.5 \times 10^9/L$ (reference range: $4.0-10.8 \times 10^9/L$)
2. Lymphocytes: $6.6 \times 10^9/L$ (reference range: $0.7-4.2 \times 10^9/L$)
3. Monocytes: $1.9 \times 10^9/L$ (reference range: $0.5-0.8 \times 10^9/L$)
4. Basophils: $0.5 \times 10^9/L$ (reference range: $0.0-0.2 \times 10^9/L$)
5. Granulocytes: $21.5 \times 10^9/L$ (reference range: $2.4-7.9 \times 10^9/L$)
6. Eosinophils: $1.0 \times 10^9/L$ (reference range: $0.0-0.7 \times 10^9/L$)

The infant was formula fed and his progress appeared to be satisfactory. His weight steadily increased to 16 lb 11oz at the age of four months. There was, however, little or no change in the lymphocytosis and monocytosis which had been recorded at birth. Over the course of the next year the child suffered several bouts of fever, cough and lethargy for which he was treated with antibiotics.

At the age of two years his mother's partner who was looking after him went to prepare a bath and left the room. He heard a thud and on returning to the room found the child on the floor apnoeic and having a seizure. The EMS was called and when they arrived they found him listless and floppy.

He was intubated and taken to the local hospital.

Physical examination showed he had numerous bruises about the front of his neck and others scattered about his body. A computerised tomography (CT) scan showed cerebral oedema consistent with cerebral anoxia and an ophthalmoscopic examination showed bilateral retinal haemorrhages. A skeletal X-Ray showed a metaphyseal fracture of the left distal humerus and a periosteal reaction of the left ulnar and left scapular.

Laboratory investigations showed:

1. PT: 18.7 seconds (reference range: 10.8-13.7 seconds)
2. INR: 1.43 (reference range: 0.9-1.1)
3. Haemoglobin: 11.0 g/dL (reference range: 13.4-16.7 g/dL)
4. Lymphocytes: $6.2 \times 10^9/L$ (reference range: $0.7-4.2 \times 10^9/L$)
5. Monocytes: $0.9 \times 10^9/L$ (reference range: $0.5-0.8 \times 10^9/L$)
6. Granulocytes: $8.5 \times 10^9/L$ (reference range: $2.4-7.9 \times 10^9/L$)

Toxic granulation and atypical lymphocytes present. The child died soon after admission and his organs were harvested for donation. At the autopsy a small subdural hematoma was found. Death was attributed to homicide.

Case #2

The 24-year-old mother had an uneventful pregnancy followed by premature rupture of the membranes, a temperature of 100°F, oligohydramnios and failure of labour to progress. A caesarean section was performed and an 8 lb 5 oz female infant was delivered and breathed and cried spontaneously. The infant was initially breast fed and was given an injection of 1 mg vitamin K before being discharged home.

At home the baby was formula fed on Enfamil by the father when the mother returned to work. The infant's progress was satisfactory until, at the age of three weeks, she suddenly developed a bout of diarrhoea

and vomiting. She was admitted to hospital where a diagnosis of gastroenteritis was made, and she was treated and discharged after a stay of three days.

When six weeks old and being fed by the father she suddenly stopped breathing, her eyes rolled back, her body stiffened and she became cyanosed. En route to hospital she was observed to have a seizure.

A CT scan was read as negative. The infant was discharged home after a stay of three days under observation. A diagnosis of seizure disorder and anaemia was made when her hemoglobin level was found to be 8.4 g/dL.

At 3 pm that same day she had another seizure and was returned to hospital where it was found she was apnoeic, hypotonic and unresponsive with a bulging anterior fontanel. She was immediately intubated and oxygenated while other investigations were carried out.

Laboratory investigations showed:

1. PT: 20.0 seconds (reference range: 11-13 seconds)
2. APTT: 100.0 seconds (reference range: 25-36 seconds)
3. Fibrinogen: 34 mg/dL (reference range: 200-400 mg/dL)
4. ALT: 107 U/L (reference range: 4-40 U/L)
5. AST: 131 U/L (reference range: 5-40 U/L)
6. GGT: 143 U/L (reference range: 7-37 U/L)
7. Alkaline Phosphatase: 274 U/L (reference range: 115-450 U/L)
8. Calcium: 8.9 mg/dL (reference range: 8-11 mg/dL)
9. Phosphorus: 2.0 mg/dL (reference range: 4-6 mg/dL)
10. Albumin: 3.0 g/dL (reference range: 3.2-4.8)

A CT Scan of the head showed "blood within the subarachnoid or subdural space consistent with recent head trauma." A healing fracture of the posterior seventh rib was present. The child died shortly after admission.

The autopsy report noted the following: A purple contusion and ecchymosis below the right ear, the scalp tissues were swollen and nine separate contusions were scat-

tered near the midline of the back; subdural hemorrhages were present over both cerebral convexities and the brain diffusely and symmetrically swollen; and a callus was seen on the left eighth rib and the body of the vertebra T10 has a recent fracture without evidence of healing. The cause of death was attributed to blunt head trauma.

Case #3

The pregnancy was complicated by discordant growth of twins and the perinatologist recommended the pregnancy be terminated at 32 weeks gestation.

The neonatal record noted that the presentation was vertex and the delivery normal. The infant cried spontaneously, but was "dusky" and was transferred to the neonatal intensive care unit where assisted respiration was instituted and continued for several days. He was discharged after a stay of five weeks.

The discharge summary noted:

1. Prematurity: 32-week small for gestational age
2. Respiratory distress: requiring additional oxygen therapy
3. Suspected sepsis
4. Necrotizing enterocolitis: associated with bloody stools necessitating antibiotic therapy and a blood transfusion
5. Hyperbilirubinaemia
6. Feeding problems
7. Thrombocytopenia
8. Anaemia of prematurity

Hepatitis B vaccine was administered the day before his discharge. There was no mention of him being given vitamin K either by mouth or by injection.

His progress appeared to be satisfactory and he was immunized with DTaP, Polio (IPV), Hemophilus Influenza, and Prevnar. He developed thrush and was treated with an oral medication. Following his vaccination, the parents noticed a change in that he appeared to be more irritable and the mother, thinking he was hungry, placed him down on a pillow and went to warm a bottle of milk for him. On returning she found him gasping for breath and he was limp and unresponsive.

The mother called 911 and the child was admitted to hospital where a cranial CT scan showed a combination of acute subdural and subarachnoid haemorrhage. Ophthalmoscope examination was not reported.

Laboratory investigations showed:

1. PT: 19.1 seconds (reference range: 14.6-16.9 seconds)
2. AST: 79 U/L (reference range: 15-37 U/L)
3. Serum protein: 5.2 g/dL (reference range: 5.4-7.0 g/dL)

The child died shortly after admission. Death was attributed to non-accidental injury.

Discussion

All three cases had an apparent life-threatening event (ALTE). One case had the classical "triad" of subdural and retinal haemorrhages with encephalopathy, and all three cases had the associated laboratory findings of increased PT indicative of vitamin K deficiency, anaemia indicative of vitamin D deficiency²⁴ and reduced serum albumin indicative of a nutritional deficiency including vitamin C deficiency. Increased AST signified disordered liver function. These are the essential clinical and laboratory features of the metabolic disorder.

Vitamin K, a fat soluble vitamin, is a cofactor for an enzymatic conversion of glutamic acid to gamma-carboxyglutamic acid by gamma-glutamyl-carboxylase, a process essential for both the clotting of blood and the mineralization of bone.²⁵ Vitamin D, also a fat soluble vitamin, is necessary for the stimulation of osteoblastic activity and the formation of bone matrix while vitamin C is essential for the synthesis of collagen which is the principal component of the skin, blood vessels, bone matrix, dentine and a deficiency causes skin lesions, dental problems, fractures and haemorrhages in scurvy.²⁶

The level of vitamin C in the blood was not determined in these children, but the fact that there was evidence of malabsorption/malnutrition suggests that a deficiency was possible.

The subdural and retinal haemorrhages,

encephalopathy, bruises and fractures in children which have hitherto been attributed to SBS, non-accidental injury, abusive head trauma or IBI are, it is suggested, the result of a metabolic disorder involving essential nutrients including vitamins C, K and D.

While the ALTE was the initial overt manifestation of the disorder it is clear that malabsorption and liver disease preceded and caused the ALTE.

In Case #1 the blood tests suggest congenital infectious mononucleosis or cytomegalovirus infection. Either would account for the severe coagulopathy precipitated by hepatic failure as shown by the alteration in the PT, the serum protein level and liver enzymes. Congenital infectious mononucleosis is known to be associated with other congenital defects,²⁷ and this child had a congenital hydrocele.

The CT scan of the head showed evidence of cerebral oedema, and an ophthalmoscopic examination showed retinal haemorrhages together with a metaphyseal fracture of the medial left distal humerus, and a periosteal reaction of the left ulnar and left scapular mistakenly suggested non-accidental injury to the attending physicians, whereas vitamin K and C deficiency would account for all the lesions.

In Case #2 the cause of the liver failure, as shown by the liver function tests, was probably the result of the infection which caused the diarrhoea and vomiting. Malnutrition is evidenced by the reduced levels of calcium, phosphorus and albumin.

In Case #3 immaturity of the liver from premature birth was most likely the event which initiated the metabolic disorder, but there is also the possibility that his vaccination may have contributed to the metabolic disorder as reported by Kalokerinos.²⁸

In the light of what is now known, the diagnoses SBS, abusive head trauma, IBI or non-accidental head trauma, are unacceptable if nutritional deficiencies have not specifically been excluded.

In a recent case the Dublin city coroner,²⁹ ignoring the opinions of the specialists involved, recorded death by natural causes

stating: "There is no evidence of cerebral trauma or 'Shaken Baby Syndrome' despite the radiological and clinical findings of subdural haemorrhage and retinal haemorrhages." Referring to the use of orthodox medical evidence, at the retrial of a woman whose life sentence was overturned having served three years for the alleged murder of a child in her care, Lord Justice Toulson³⁰ said: "Today's orthodoxy may become tomorrow's outdated learning."

Conclusion

The contemporary notions of retinal and intracerebral haemorrhage being caused by rotation and slamming the head of the infant against a hard object is not validated by science. When one considers the evidence of nutritional deficiencies and liver dysfunction in all three cases shown here, it is entirely conceivable that a metabolic disorder involving vitamins C, K and D is the main cause of the bruising, bleeding and fractures seen in these children.

Competing Interests

The author has given evidence for the defence in courts in England, United States of America and Australia, and has received payment for these services

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