

Dr Macdonald22/2/05  
ep

re: Claire Roberts (deceased)

I have had sight of the medical notes for Claire Roberts, having been asked to comment on her attendance to the Royal Belfast Hospital for Sick Children on 21st October 1996. I understand that Dr Robert Bingham, consultant paediatric anaesthetist at the Hospital for Sick Children, Great Ormond Street, London has been asked to comment on the fluid management and the role that Claire Robert's hyponatraemia played in her subsequent demise.

I will outline the sequence of events from the medical notes and comment on her management.

The sequence of events in Claire Roberts' life from 21<sup>st</sup> October 1996

GENERAL PRACTITIONER

Claire Roberts had been referred to the Accident and Emergency Department by the General Practitioner with the following history:

9 year old girl with severe learning difficulties and a past medical history of epilepsy. He noted that she had been fit free for 3 years and had been weaned off her anti-epileptic medication eighteen months previously. She had not been speaking since coming home from school, where she had been noted as being very lethargic. She had vomited 3 times. Speech slurred earlier on.

The general practitioner noted on examination that she was pale, that her pupils were reacting but that she did not like light. There was no neck stiffness, no raised temperature but increased tone on the right side of her body, and the right plantar reflex was up going (*which is abnormal*). Her ear, nose and throat examination was normal. Her respiratory system was clear.

Correctly, the general practitioner was concerned about her neurological status and made a provisional diagnosis of further fits or underlying infection (*probably questioning the possibility of encephalitis, that is an inflammation of the brain*).

INITIAL ASSESSMENT AT ROYAL BELFAST HOSPITAL FOR SICK CHILDREN

She attended the Children's Hospital with her mother at 18.57 hours and was seen at 19.15 hours.

~~Her vital signs were:~~ respiratory rate 24 breaths per minute, heart rate 96 beats per minute, which are both elevated for her age. She did not have a raised temperature.

The history and examination were largely unchanged from examination made by the GP. Her speech was noted to be very slurred and that she was hardly speaking. She was drowsy and the neurological examination was abnormal in that the right side of her body and there was an increased tone noted. There were bilateral down going plantars i.e. now both right and left plantar reflexes were abnormal. There was clonus in both ankles, which is indicative of abnormal neurological functioning.

The decision to admit her was made with a provisional diagnosis of ? encephalitis, signed by Dr O'Hare.

#### INITIAL ADMISSION NOTES

Claire was admitted to Allan ward.

8pm The first medical notes of her admission were made at 8 pm.

She was noted to be sitting up and staring vacantly, only responding intermittently to voice, but responded to pain. A full examination could not be conducted owing to her inability to co-operate. She ascribed a preliminary diagnosis of viral infection. I note that encephalitis had been scoured out of the notes.

Appropriate investigations were instigated and she was managed by being commenced on intravenous fluids and in the notes intravenous diazepam prescribed if she were to show seizure activity. She was to be reassessed after the intravenous fluids had been given. This is signed by Dr O'Hare.

At midnight she was seen by the SHO who recorded that she was slightly more responsive and that she did not have the clinical features of meningitis.

The sodium is 132 mmol/l which is on the low side of normal. She had a raised white cell count which may be indicative of infection.

The next day, on the ward round was lead by Dr Sands. Claire had not had any seizure activity and showed little response as compared with her normal status.

On examination her pupils were sluggish in response to light stimulus and she showed abnormal long tract signs (that is, abnormal neurological responses).

From these findings, Dr Sands made the differential diagnoses of non-fitting status, encephalitis or encephalopathy and it was noted that she should have diazepam which is part of the treatment for the control of seizures. Dr Webb, consultant neurologist was contacted. Dr Gaston who was the consultant in charge of her long term management previously was to be contacted.

#### ~~DR WEBB~~

Dr Webb subsequently wrote in the notes at 4 pm on 22<sup>nd</sup> October 1996 that he found that she did not have any features of meningitis, that she was rousable to voice, that she withdrew from power stimuli, that she had clonus at both ankles, had increased tone and that she was sitting up staring vacantly and not obeying commands.

'I don't have a clear picture of the prodrome and yesterdays episodes. Her motor findings today are probably long standing but this needs to be checked with notes. The picture is of acute encephalopathy most probably postictal in nature.'

Dr Webb suggested commencing more antiepileptic medication, hourly neurological recording and for her to have a CT the next day should she not 'wake up'.

She was noted not to have responded to the antiepileptic medication and therefore additional medication was commenced.

Dr Webb reviewed Claire at 17:00, noting that she had had the loading dose of the anti-epileptic medication, phenytoin (*initially this drug is given over a period of time, as a large dose so that satisfactory blood levels are achieved*) and a dose of midazolam which is also used in the treatment of seizures.

He noted that she continued to appear to be 'largely unresponsive', that she responded to deep pain by flexing her left arm and had a facial grimace. There was no vocalisation noted.

He obtained further history of the stiffening on the right hand side of the body being present on Monday with focal signs. There was contact with a cousin who had had gastrointestinal upset the preceding day.

Dr Webb prescribed antibiotic and anti viral medication to be started as a precaution albeit he thought the likelihood for either a bacterial or viral meningitis to be present was low; he asked for viral cultures to be taken to see if a viral infection could account for Claire's condition and that another anti-epileptic medication be started.

The notes record that at 23.30, the results of the blood samples were available, showing hyponatraemia and her fluid management was altered. I will defer to Dr Bingham regarding the management of her fluid regime.

At about 3 am on the 23<sup>rd</sup> October 1996, she had a respiratory arrest and developed fixed dilated pupils. She was intubated and transferred to the paediatric intensive care unit.

At 4am her examination revealed fixed dilated pupils, with bilateral papilloedema (*suggesting raised intracranial pressure, i.e. the pressure within her brain was elevated*), and that she was no longer responsive to painful stimuli.

She was reviewed at 4.30 am by Dr Webb, who considered her to have the syndrome of inappropriate anti-diuretic hormone production, leading to hyponatraemia and cerebral oedema. He also thought that she had coned (*that is her brain due to swelling had become forced down the main outlet within the skull, causing irreparable damage to the brain*), following prolonged seizure activity. She had had mannitol, medication which is used to decrease brain swelling by drawing out fluid by virtual of its osmotic effects, but that this had had no effect on the pupillary size, and that there were no eye moments. This composite picture is alarming and suggested that brain death had occurred.

Dr Webb requested an urgent CT of Claire's head which confirmed the clinical findings of diffuse swelling of the brain, without any focal abnormality being seen (recorded in the notes at 5 am on 23<sup>rd</sup> October 1996).

At 6 am the brain stem death criteria were applied by Dr Webb.

The laboratory sample at the time of brain stem testing recorded the sodium level as 129 mmol/l.

These brain stem test criteria were repeated and confirmed at 18:25.

Intensive care support was withdrawn from Claire at 18:45 and a death certificate for cerebral oedema secondary to status epilepticus was written.

## CONCLUSIONS

Claire Roberts was admitted with abnormal neurological symptoms and signs. The diagnosis of encephalitis/ encephalopathy was made at an early stage of her admission and measures taken to treat the likely diagnosis of non-convulsive epilepsy. There was a background of seizure activity in her past medical history and hence the probability of this diagnosis was high, given her presentation.

Dr Webb played a key part in the management of Claire and his initial assessment was considered and defined a clear management plan that incorporated treatment of meningitis (although as he rightly said, this was an unlikely diagnosis, however, it was essential to instigate treatment for meningitis. Failure to do so would lead to permanent disability and a possible death if meningitis had been present).

The underlying diagnosis was that of non-convulsive status epilepticus; a condition where the electrical activity of the brain is disrupted but there are not the somatic manifestations of seizure activity apparent such as is seen with other forms of fitting such as jerking convulsions. If untreated non-convulsive status epilepticus can lead to worsen any disability and be a cause of death. The term status meaning that the fitting is established (e.g. one definition for status epilepsy is that it has lasted for more than 20 minutes).

The management plan to treat the possibility of non-convulsive status epilepticus was correct at the time of practice.

Claire Robert's subsequent management was correct and her course of management on the ward and PICU was appropriate. The parents were invited to return for further discussions after the withdrawal of treatment.

I have not commented upon the hyponatraemia which has been addressed by Dr Bingham. Claire Robert's treatment from the point of view of management of her neurological presentation was considered, and Dr Webb and other members of the team looking after Claire gave careful and informed advice.