

**BRIEF FOR EXPERT ON PAEDIATRIC NEUROLOGY
ADAM STRAIN**

Introduction

1. Adam Strain is one of 4 children who are the subject of a public inquiry being conducted by John O’Hara QC.
2. Adam was born on 4th August 1991. He died on 28th November 1995 in the Royal Belfast Hospital for Sick Children (“RBHSC”) following kidney transplant surgery. The Inquest into his death was conducted on 18th and 21st June 1996 by John Leckey the Coroner for Greater Belfast, who engaged as experts: (i) Dr. Edward Sumner then Consultant Paediatric Anaesthetist at Great Ormond Street Hospital for Sick Children (“Great Ormond Street”); (ii) Dr. John Alexander Consultant Anaesthetist at Belfast City Hospital; and (iii) Professor Peter Berry of the Department of Paediatric Pathology in St. Michael’s Hospital, Bristol. The Inquest Verdict identified Cerebral Oedema as the cause of his death with Dilutional Hyponatraemia as a contributory factor.
3. The other 3 children are :
 - (1) Claire Roberts was born on 10th January 1987. She was admitted to the RBHSC on 21st October 1996 with a history of malaise, vomiting and drowsiness and she died on 23rd October 1996. Her medical certificate recorded the cause of her death as Cerebral Oedema and Status Epilepticus. That certification was subsequently challenged after a television documentary into the deaths of Adam and 2 other children (Lucy Crawford and Raychel Ferguson).

The Inquest into Claire’s death was carried out by John Leckey on 4th May 2006 who engaged as experts Dr. Robert Bingham (Consultant Paediatric Anaesthetist at Great Ormond Street) and Dr. Ian Maconochie (Consultant in Paediatric A&E Medicine at St Mary’s, London). The Inquest Verdict found the cause of Claire’s death to be Cerebral Oedema with Hyponatraemia as a contributory factor.

- (2) Raychel Ferguson was born on 4th February 1992. She was admitted to the Altnagelvin Area Hospital on 7th June 2001 with suspected appendicitis. An appendectomy was performed on 8th June 2001. She was transferred to the RBHSC on 9th June 2001 where brain stem tests were shown to be negative and she was pronounced dead on 10th June 2001. The Autopsy Report dated 11th June 2001 concluded that the cause of her death was Cerebral Oedema caused by Hyponatraemia.

The Inquest into Raychel's death was conducted on 5th February 2003 by John Leckey who once more engaged Dr. Edward Sumner as an expert. The Inquest Verdict found the cause of Raychel's death to be Cerebral Oedema with Acute Dilutional Hyponatraemia as a contributory factor. It also made findings that the Hyponatraemia was caused by a combination of inadequate electrolyte replacement following severe post-operative vomiting and water retention resulting from the secretion of anti-diuretic hormone (ADH).

- (3) Conor Mitchell was born on 12th October 1987 with cerebral palsy. He was admitted to A&E Craigavon Hospital on 8th May 2003 with signs of dehydration and for observation. He was transferred to the RBHSC on 9th May 2003 where brain stem tests were shown to be negative and he was pronounced dead on 12th May 2003.

The Inquest into Conor's death was conducted on 9th June 2004 by John Leckey, Coroner who again engaged Dr. Edward Sumner as an expert. Despite the Inquest, the precise cause of Conor's death remains unclear.

The clinical diagnosis of Dr. Janice Bothwell (Paediatric Consultant) at the RBHSC was brainstem dysfunction with Cerebral Oedema related to viral illness, over-rehydration/inappropriate fluid management and status epilepticus causing hypoxia. Dr. Brian Herron from the Department of Neuropathy, Institute of Pathology, Belfast performed the autopsy. He was unsure what 'sparked off' the seizure activity and the extent to which it contributed to the swelling of Conor's brain but he considered that the major hypernatraemia occurred after brainstem death and therefore probably played no part in the cause of the brain swelling. He concluded that the ultimate cause of death was Cerebral Oedema.

Dr. Edward Sumner commented in his Report of November 2003 that Conor died of the acute effects of cerebral swelling which caused coning and brainstem death but he remained uncertain why. He noted that the volume of intravenous fluids was not excessive and the type appropriate but queried the initial rate of administration. That query was raised in his correspondence shortly after the Inquest Verdict. In that correspondence, Dr. Sumner described the fluid management regime as 'sub-optimal'.

The Inquest Verdict stated the cause of death to be Brainstem Failure with Cerebral Oedema, Hypoxia, Ischemia, Seizures and Infarction and Cerebral Palsy as contributing factors.

4. The impetus for this Inquiry was a UTV Live 'Insight' documentary 'When Hospitals Kill' shown on 21st October 2004. The documentary primarily focused on the death of a toddler called Lucy (who died in hospital in 2000 and whose death was subsequently found to have been as a result of hyponatraemia). The

programme makers identified what they considered to have been significant shortcomings of personnel at the Erne Hospital where Lucy had been initially treated before being transferred to the RBHSC. In effect, the programme alleged a cover-up and it criticised the hospital, the Trust and the Chief Medical Officer. The programme also referred to the deaths of Adam and Raychel in which hyponatraemia had similarly played a part. At that time, no connection had been made with the deaths of Claire and Conor.

Original Terms of Reference

5. On 1st November 2004 by Angela Smith MP who was the Minister with responsibility for the Department of Health, Social Services and Public Safety. The Inquiry was established under the Health and Personal Social Services (Northern Ireland) Order 1972, under the powers conferred on the Department by Article 54 and Schedule 8.¹

Changes

6. The work of the Inquiry was suspended in October 2005 due to investigations by the Police Service of Northern Ireland (PSNI) into the deaths of the 3 children. It was decided not to prosecute anyone in relation to the children's deaths.
7. The Inquiry re-started its work in 2008 but with a number of significant changes. Firstly, Lucy's family wished to have her excluded from the Inquiry's work for personal reasons. The Minister therefore issued the following Revised Terms of Reference to the Inquiry, which govern the work of the Inquiry:

1. The care and treatment of Adam Strain and Raychel Ferguson, especially in relation to the management of fluid balance and the choice and administration of intravenous fluids in each case.

2. The actions of the statutory authorities, other organisations and responsible individuals concerned in the procedures, investigations and events which followed the deaths of Adam Strain and Raychel Ferguson.

3. The communications with and explanations given to the respective families and others by the relevant authorities.

In addition, Mr O'Hara will:

- (a) Report by 1 June 2005 or such date as may be agreed with the Department, on the areas specifically identified above and, at his discretion, examine and report on any other matters which arise in connection with the Inquiry.

- (b) Make such recommendations to the Department of Health, Social services and Public Safety as he considers necessary and appropriate.

¹ The legislation currently governing the Inquiry is Health and Personal Social Services (Northern Ireland) Order 1972 and continues by virtue of section 23 of the Interpretation Act (Northern Ireland) 1954 (as amended).

8. Secondly, Claire and Conor were included into the Inquiry's work by the Chairman. In Claire's case that decision arose out of an acknowledgement by the RBHSC at the end of 2004 that hyponatraemia had played a part in Claire's death. In Conor's case, the decision arose out of apparent fluid mismanagement in his care soon after the implementation of Guidelines on Hyponatraemia issued by the Chief Medical Officer which stressed the importance of fluid management.

9. The effect of the Revised Terms of Reference was to exclude all explicit references to Lucy. The Chairman interpreted them as:

... the terms still permit and indeed require an investigation into the events which followed Lucy's death such as the failure to identify the correct cause of death and the alleged Sperrin Lakeland cover-up because they contributed, arguably, to the death of Raychel in Altnagelvin. This reflects the contention that had the circumstances of Lucy's death been identified correctly and had lessons been learned from the way in which fluids were administered to her, defective fluid management would not have occurred so soon afterwards (only 14 months later) in Altnagelvin, a hospital within the same Western Health and Social Services Board area.

10. Claire's case is being investigated in accordance with precisely the same terms as those of Adam and Raychel.

11. The investigation of Conor will address more limited issues since hyponatraemia was not thought to be a cause of his death (if anything he developed hypernatraemia). Also, the fluid mismanagement referred to by Dr. Sumner was not considered to have caused his death. The Chairman has stated:

"It is obviously a matter of concern if guidelines which have been introduced as a result of a previous death or deaths and which are aimed at avoiding similar events in the future, are not properly communicated to hospital staff and followed. It is relevant to the investigation to be conducted by the Inquiry whether and to what extent the guidelines had been disseminated and followed in the period since they were published. Another matter of interest is whether the fact that Conor was being treated on an adult ward rather than a children's ward made any difference to the way in which it appears that the guidelines may not have been followed.

Accordingly, the Inquiry will investigate the way in which the guidelines had been circulated by the Department, the way in which they had been made known to hospital staff and the steps, if any, which had been taken to ensure that they were being followed. While this is an issue of general importance, it will be informed by an examination of the way in which the guidelines had been introduced and followed in Craigavon Area Hospital by May 2003."

Role of the Experts

12. The Role of the Experts to the Inquiry is set out in 'Protocol No.4: Experts', a copy of which is attached.

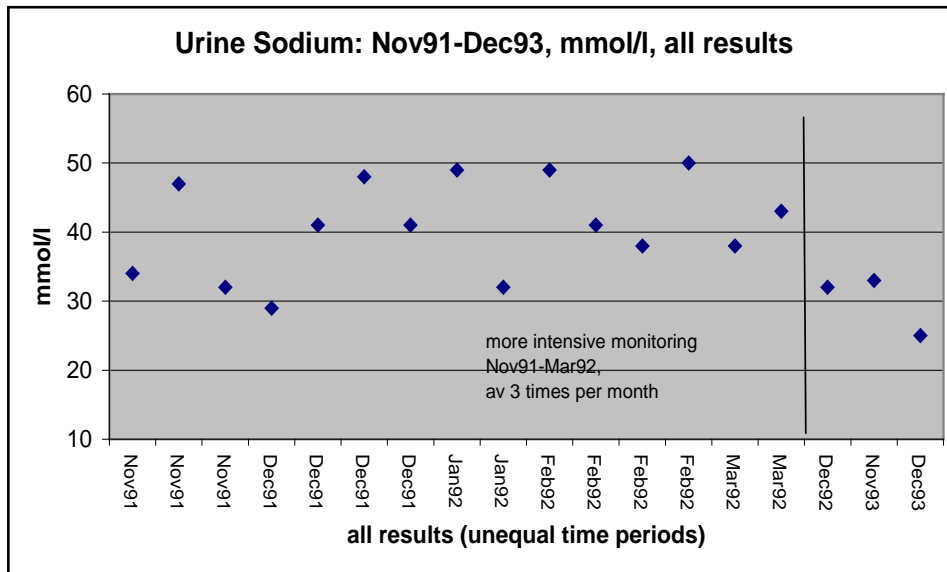
13. In particular the Inquiry engages Experts on a case-by-case basis as Expert Witnesses. In addition to your expertise as an expert on paediatric neurology, the Inquiry has also appointed Experts in Adam's case on Transplant Surgery, Paediatric Nephrology, Hyponatraemia, Paediatric Nursing, Paediatric Anaesthetics, Radiology, and Paediatric Neuropathology.²
14. Like all of those other Experts the Inquiry will need you to provide your Expert opinion in the form of a Report incorporating an 'expert declaration' and attached to a Witness Statement. Your Report will be made public and you may be required to attend the Oral Hearings and present your views.

Background to Adam

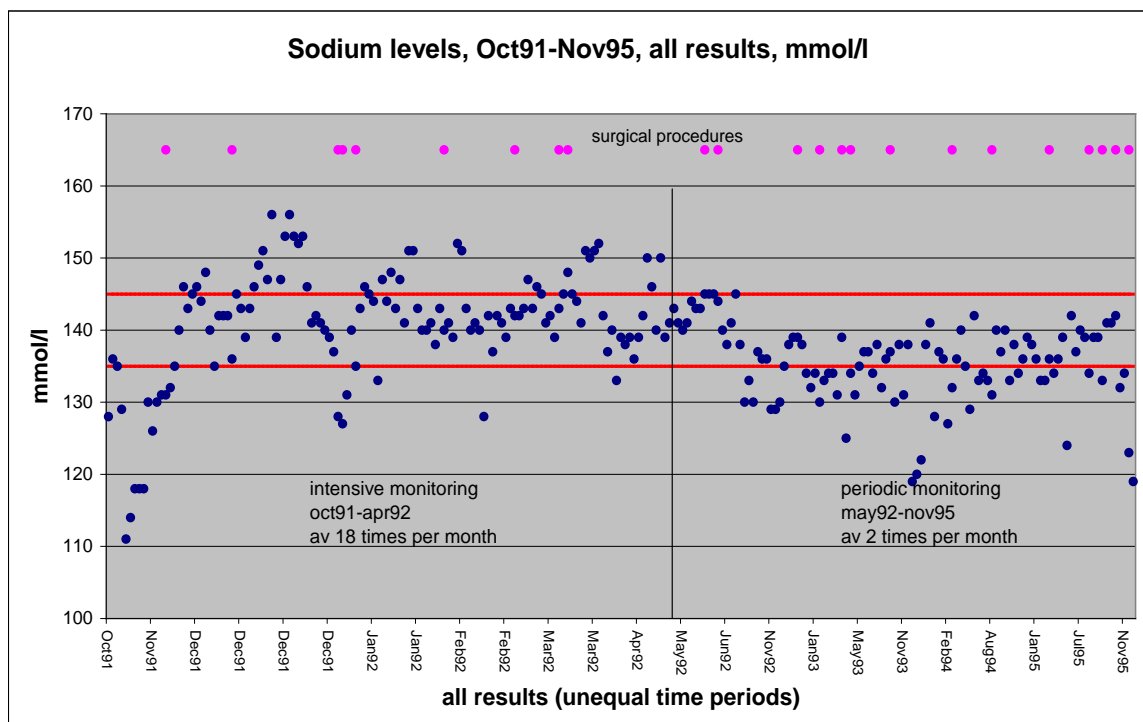
15. Adam was born with cystic dysplastic kidneys. He developed problems with the drainage of his kidneys related to obstruction and vesico-ureteric reflux. He was referred to the RBHSC from the Ulster Hospital in Dundonald when he was a few months old and came under the care of Dr. Maurice Savage (Consultant Paediatric Nephrologist)³ and Mr. Stephen Brown (Consultant Paediatric Surgeon).
16. Thereafter, Adam had multiple operations to his urinary tract during which he was largely under the care of Mr. Stephen Brown. He had re-implantation of his ureters on 2 occasions and had nephrostomies, which were performed during the early months of his life. On several occasions, he was critically ill and required care in PICU and a brief period of dialysis due to acute renal failure. In addition a fundoplication procedure was carried out in 1992 when Adam was less than a year old, to help prevent gastro-oesophageal reflux. Eventually he required all his nutrition through a gastrostomy tube. In 1993 he had a cystoscopy and PEG gastrostomy. The PEG was changed in October 1995.
17. Adam was subject to recurrent urinary tract infections and his renal function deteriorated to the point where, in 1994, he required dialysis for uraemia. His mother was trained in the home peritoneal dialysis technique so that he could be dialysed at home. His urine output was quite large but of poor quality and he was described as being polyuric. Biochemistry tests carried out when he was a few months old showed the sodium content of his urine to be 29 - 52 mmol/l. A graph of all Adam's recorded urine sodium results is shown below:

² Dr. Malcolm Coulthard (Paediatric Nephrology); Professor Peter Gross (Hyponatraemia); Ms. Sally Ramsay (Paediatric Nursing); Professor John Forsyth & Mr. Keith Rigg (Transplant Surgeons); Dr. Simon Haynes (Paediatric Anaesthetics); Dr. Caren Landes (Radiology); Professor Fenella Kirkham (Paediatric Neurology); Dr. Wayne Squier (Neuropathology); Professor Sebastian Lucas (Pathology); Professor Aidan Mullan (Clinical Governance); Mr. Stephen Ramsden (Hospital Management & Governance)

³ Now Professor Maurice Savage



18. According to his nephrologist, Dr. Maurice Savage, Adam had a potential for hyponatraemia and he received sodium supplements in his feeds. A graph of all of his recorded blood sodium levels is shown below with the parallel lines indicating the normal range of 135-145mmol/l:



19. The management of his serum sodium levels appears to have been largely carried out under the care of Messrs. Victor Boston and Stephen Brown, both Consultant Paediatric Surgeons. Despite that, his recorded sodium levels for 1995, the year of his transplant surgery, show one very low result of 124 mmol/l

and a number below the normal range of 135-145 mmol/l. Furthermore, in Adam's first year of life his recorded sodium levels fell as low as 111 mmol/l, 114 mmol/l and 118 mmol/l. Thereafter there were numerous occasions when his recorded serum sodium levels fell below the normal range.

20. Adam was put on call for a kidney transplant once he was placed on dialysis. His tube feeds in the months prior to the transplantation surgery were slightly over 2 litres per day and he passed in excess of 1 litre of urine each day.
21. Adam received the offer of a reasonably matched kidney on 26th November 1995. The donor kidney had been removed in Glasgow from a heart-beating 16-year-old donor with normal renal function at 01:42 on 26th November 1995. Adam's Transplant surgery was scheduled for 06:00 on 27th November 1995, which was subsequently put back to 07:00.
22. At 23:00 on 26th November 1995, Adam's serum sodium was recorded as 139 mmol/l and Hb 10.5 gm/dl. As part of the preparation for his surgery, his feeds were changed although there remains an issue as to exactly what they were changed to. According to his charts, he was given 952 ml of 'clear fluid' to stop 2 hours before going into theatre. The nursing records do not state the nature of the 'clear fluids' given. Some witnesses have claimed that fluid was Dioralyte (containing 60 mmol of sodium chloride/L). However, Dr. Maurice Savage corrected his Deposition to the Coroner to delete 'Dioralyte' and substitute 'N/S Saline Dextrose'. In any event, it is thought that he received just over 1 litre of fluids. Apparently, it was planned between Dr. Maurice Savage and Dr. Robert Taylor (Consultant Paediatric Anaesthetist) that Adam should receive intravenous fluid (75 ml/h) after the tube feeds were discontinued and have his blood chemistry checked before going to theatre. It seems that those checks did not take place. Although the Inquiry has very recently been provided with a laboratory result of a blood test taken some time on 26th November 1995 that measures Adam's serum sodium level at 133 mmol/L. Adam's notes do not record the time at which the sample was taken nor the receipt of such a result.
23. Once again, the clinicians have provided different explanations as to why, so far as they are concerned, the envisaged checks were not undertaken. On one basis, it was because it proved difficult to achieve venous access, whilst on another it was because of the potential delay in receiving results back from the laboratory and that it was not considered to be a priority.
24. The main events surrounding Adam's pre-operative, peri-operative and post-operative care and treatment are summarised in the attached chronology. As can be seen from that chronology, Adam failed to wake at the end of surgery and at about 12:00 noon his pupils were found to be fixed and dilated.
25. An emergency CT scan of Adam's brain was carried out at 13:15. Dr. Charles McKinstry (Consultant Neuroradiologist at the Royal) wrote up the results of it

in Adam's notes⁴ as: *"There is marked generalised cerebral swelling with ... of the lateral ventricles and obliteration of the third ... ventricles, basal cisterns and ... sulci. No focal abnormality seen"*. The subsequent report states:⁵ *"The lateral ventricles are very small. The third ventricle is not clearly seen. There is no mid-line shift. The brain appears swollen. No focal parenchymal lesion can be seen. Conclusion: These appearances are in keeping with diffuse brain oedema"*.

26. Dr. David Webb (Consultant Paediatric Nephrologist) examined Adam at 19:30 that evening and carried out a first brain stem test. He concluded that the results fulfilled the criteria for preliminary confirmation of brain stem death. He noted *"severe extensive bilateral fundal haemorrhages suggestive of acute raised intracranial pressure"*.⁶ He also reviewed Adam's CT which he described as showing: *"diffused generalised cerebral oedema with obliteration of the basal cisterns fulfilling the radiological criteria for coning"*.⁷
27. A pre-transplant surgery chest x-ray was required in accordance with the RBHSC Protocol on 'Renal Transplantation in Small Children'. Dr. Donagh O'Neill (SHO in paediatrics) requested the chest x-ray (Ref: 057-019-028) but it is unclear whether that actually happened. Nevertheless, the entry in Adam's medical notes records that his chest was clear on the evening of 26th November 1995.⁸ There were 2 post-transplant surgery chest x-rays. The first was timed at 13:20 on 27th November 1995 (Ref: 058-035-140), whilst the second was timed that evening at 21:30 (Ref: 058-035-142). It is recorded in Adam's medical notes that the first showed some pulmonary oedema⁹ and the second showed that there was still quite marked pulmonary oedema with fluid in horizontal fissure.¹⁰
28. A post-mortem was carried out on 29th November 1995 by Dr. Armour who reported the cause of Adam's death as: 1(a) cerebral oedema due to (b) dilutional hyponatraemia and impaired cerebral perfusion during renal transplant.
29. For the purposes of her Report and to assist the Coroner, Dr. Armour sought a second opinion on the histological slides from Professor Jeremy Berry (Professor of Paediatric Pathology). He was sent a request for an opinion on 22nd December 1995, in which she stated that: *"At post-mortem I found gross cerebral oedema (1,320 gms). The brain and spinal cord are fixing and a neuropathological opinion will be requested"*.¹¹ Professor Berry was provided with slides of: (i) the native kidneys

⁴ Ref: 058-035-138 - attached as part of Adam's medical notes and records. See also his Inquiry Witness Statement, Ref: WS-111/1 - attached

⁵ Ref: 301-081 - attached

⁶ Ref: 059-061-147 - attached

⁷ Ref: 059-061-147 - attached

⁸ Ref: 058-035-131 - attached as part of Adam's medical notes and records

⁹ Ref: 058-035-140 - attached

¹⁰ Ref: 058-035-142 - attached

¹¹ Ref: 011-029-151 - attached

- and the donor kidney; (ii) spleen; (iii) lungs; (iv) liver; (v) lymphnode and was given the un-fixed brain weight as 1,320g. He noted that there was unexplained cellular change in the hepatocytes scattered throughout his liver but he did not know the significance of it. He concluded that the transplanted kidney was infarcted at or before the time of transplantation.¹²
30. On 12th January 1996, the brain was weighed and cut after fixation. The recorded fixed weight, as shown on the Report of Autopsy, is: (a) brain: 1,680g; (b) cerebellum: 154g; and (c) brain stem: 22g. Histological slides were made and the brain was photographed sequentially. The Inquiry has not been provided with copies of the photographs. However, two photographs of Adam's brain can be seen in an article by Dr. Armour. One is described as showing "*marked uncal swelling and swelling of cerebellar tonsils*". Whilst the other is described as showing "*cut section of brain showing slit-like ventricles*".¹³
31. Apparently, Dr. Armour also showed slides of the brain and related material to Dr. Meenaskshi Mirakhur (Consultant Neuropathologist at the Royal) for a second opinion. Blocks were taken from: (i) right frontal white matter; (ii) left cingulate gyrus; (iii) left basal ganglia; (iv) right and left hippocampus; (v) left occipital lobe; (vi) cerebellum; (vii) pons in toto; (viii) thalamus and the brain was photographed sequentially. Blocks were also taken from: (i) cervical; (ii) thoracic; (iii) lumbar. There is no record of a formal request from the State Pathologist's Department and to date the Inquiry has not been provided with any note or opinion from Dr. Mirakhur.¹⁴ According to Dr. Armour's Inquiry Witness Statement,¹⁵ Dr. Mirakhur's views were consistent with her own description of and comments on the brain as stated in her Report on Autopsy¹⁶
32. It would appear that on 8th December 1995 Dr. Armour also showed "*slides etc.*" to Dr. O'Hara (Consultant Paediatric Pathologist at the RBHSC) and Dr. Bharucha (Consultant Haematologist at Belfast City Hospital). It is not clear exactly what slides were shown as the only evidence of this is a note made by the Coroner following his conversation with Dr. Armour.¹⁷ Apparently, Drs. O'Hara and Bharucha both stated: "*there was clear evidence of hypoxia/anoxia/anaphylactic [sic] reaction*". Dr. Armour's Report on Autopsy was provided on 24th April 1996. It includes no reference to the: "*clear evidence of hypoxia*" that Drs. O'Hara and Bharucha are said to have identified. Rather it states: "*There was no evidence of terminal hypoxia*".¹⁸

¹² Ref: 011-007-020 - attached

¹³ Armour - Dilutional hyponatraemia: a cause of massive fatal intraoperative cerebral oedema in a child undergoing renal transplantation, J Clin Pathol 1997; 50:444-446 - attached. The photographs of the brain (Figures 1 & 2) are also attached separately.

¹⁴ Ref: INQ-0486-11 - attached

¹⁵ Ref: WS-012/1, p.2, Q2b - attached

¹⁶ Ref: 011-010-034- attached

¹⁷ Ref: 011-025-125 - attached

¹⁸ Ref: 011-010-040- attached

33. The Report on Autopsy records the fluids given to Adam. Dr. Armour also reports and comments that the fixed weight of the brain at post-mortem was 1,680g, the average weight for a boy of this age being 1,300g and the average weight of a man's brain being 1,450g and that it was the "*effects of this massive swelling of the brain which caused his death*".¹⁹ However, in her contemporaneous notes of the autopsy, Dr Armour records the weight of the brain as 1,320g (or possibly 1,302g).²⁰
34. The Inquest that was subsequently conducted into Adam's death on 18th and 21st June 1996 recorded the Verdict that the cause of his death was:

1(A) Cerebral Oedema

due to

(B) Dilutional Hyponatraemia and impaired cerebral perfusion during renal transplant operation for chronic renal failure (congenital obstructive uropathy)

Findings:

The onset of cerebral oedema was caused by the acute onset of hyponatraemia from the excess administration of fluids containing only very small amounts of sodium and this was exacerbated by blood loss and possibly the overnight dialysis and the obstruction of the venous drainage to the head

35. The Coroner, Mr. John Leckey, was assisted in reaching that Verdict by Dr. Edward Sumner (Consultant Paediatric Anaesthetist) who was retained to prepare a Report on the circumstances of Adam's death. Dr. Sumner concluded in his Report dated 22nd January 1996:

I believe that on a balance of probabilities **Adam's gross cerebral oedema was caused by the acute onset of hyponatraemia** (see reference) from the excess administration of fluids containing only very small amounts of sodium (dextrose-saline and plasma). This state was exacerbated by the blood loss and possibly by the overnight dialysis.

A further exacerbating cause may have been the obstruction to the venous drainage of the head. If drugs such as antibiotics were administered through a venous line in a partially obstructed neck vein then it is possible that they could cause some cerebral damage as well.

(Emphasis added)

36. Dr. Sumner also gave evidence at Adam's Inquest and his Deposition of 18th June 1996 records him as having expressed the following views:

All the fluids given after dialysis may have been given to increase central venous pressure. It may have had the effect of causing the dilution of the sodium in the body. **Fluid balance in paediatrics is a more controversial area with a variety of views. With kidney transplants one gives more fluids than in other operations** [*"it is usual to be generous with fluids to maintain a CVP of 10-12 to optimise perfusion of the new kidney and to establish its urine-producing function"*²¹]. When the new kidney is perfused it is vital that sufficient fluids are available. I got the impression that Dr. Taylor was not believing the CVP readings he was getting. I

¹⁹ Ref: 011-010-040 - attached

²⁰ Ref: INQ-0319-11 - attached

²¹ See Dr. Sumner's Report of 22nd January 1996 at ref:011-011-059 - attached

believe they were probably correct but high. I think I would have believed them. A high CVP can mean too much fluid has been administered²² ... The **low sodium was indicative of the hyponatraemia. Below 128 is a hyponatraemic state.**

(Emphasis and parenthesis added)

37. Dr. Robert Taylor (Consultant Paediatric Anaesthetist) gave evidence at the Inquest. His Deposition of 21st June 1996 shows that he disagreed with Dr. Sumner's principal finding:

I cannot understand why a fluid regime employed successfully with Adam previously, led on this occasion to dilutional hyponatraemia ... I believe that **the underlying cause of the cerebral oedema was hyponatraemia (not dilutional)** during renal transplant operation.

...

Adam was the only child with polyuric renal failure I have anaesthetised for renal transplant. He needed a greater amount of fluid because of the nature of the operation [*"All the more important in this case is the need to avoid dehydration that will deprive the donor kidney of sufficient fluid to produce urine"*²³]. I believe the fluids given were neither restrictive or excessive. **The new kidney did not work leading to a re-assessment of the fluids given.** This made us think we have underestimated fluid and we gave a fluid bolus at 9.32.

(Emphasis added)

38. The circumstances of the calculation of the fluids given to Adam and the actual amounts involved (bearing in mind his 'polyuric condition'²⁴) are important issues for the Inquiry as they go to whether Adam's hyponatraemia might have been avoided by appropriate fluid management.

Requirements

39. The Inquiry requires your assistance with the following matters, arising out of the material received to date:

- (1) Is there a means of estimating for a 4 year old boy of approximately 20 kg:
 - (i) The extra-cerebral fluid space available to accommodate cerebral oedema (i.e. 'reserve capacity')? If so, please explain it and provide the estimate.

²² Dr. Sumner prepared his Report on the basis that Adam received 900mls of Dioralyte. See at ref: 011-011-055 - attached. That figure was corrected in correspondence between the Coroner and Dr. Armour but it is not clear that the correspondence from Adam's mother referring to the lower figure was passed to Dr. Sumner. Dr. Armour thought that the difference between the two figures made no difference to her opinion on the cause of Adam's death: *"It is not just the volume of fluid he received but the type."* See at ref: 011-079-214 - attached

²³ See Deposition at ref:011-014-100 - attached

²⁴ See letter dated 2nd March 1995 from Mr. Maurice Savage (Consultant Paediatric Nephrologist) to Dr. Scott (Adam's GP) explaining: *"The problem is he still needs about 2 litres a day because of his polyuric renal failure"* (Ref: 057-072-133) - attached.

- (ii) Volume of fluid and the rate at it would need to enter the brain so as to cause death due to coning from raised intracranial pressure? If so, please explain it and provide the estimate.
- (iii) Is it possible to describe for such a child the relationship between increasing intracranial volume and intracranial pressure? If so, please explain it.
- (2) If Adam's total body water had been expanded by 10 per cent over a period of 2½ hours from the induction of anaesthesia:
- (i) What effect would that then have had on his brain weight?
- (ii) If there was osmotic overload, what effect would you expect that to have on his brain?
- (iii) Is there a means of establishing (or estimating) the volume of free water intake that would be sufficient to cause fatal cerebral oedema? If so, please explain it and provide the figure.
- (3) Adam's serum sodium fell from 139 mmol/L (laboratory sample) measured preoperatively at 21:00 on 26th November 1995 (-10 hours) to 123 mmol/L (blood gas machine sample) 2½ hours after induction and 119 mmol/L (laboratory sample) 4½ hours after induction. There is also an un-timed value of 133 mmol/L (laboratory sample) of a serum sample taken some time in the evening of 26th November 1995:
- (i) Is it possible to calculate what proportion of the free water infused would have contributed to Adam's cerebral oedema and what proportion would have been diffused throughout other organs?
- (ii) If so, what are those proportions?
- (4) The transplant surgery concluded at about 11:00 on 27th November 1995. At about 12:00 noon Adam was found not to be breathing spontaneously and to have fixed and dilated pupils. There were no signs of recovery and death was pronounced about 24 hours later after two brain-stem death tests had been performed:
- (i) Is it reasonable to assume that the fatal event is likely to have occurred at or before 12:00 noon rather than after that time?
- (5) It has been suggested that a contributor to fatal cerebral oedema may have been obstruction to venous drainage from the brain. This is based on the following:
- An operative note made some years earlier that the left common facial vein had been ligated.
 - A 'Broviac line' was inserted in 1992, which might have involved accidentally putting a suture around the left internal jugular vein.

- The pathologist who conducted the autopsy stated rather *“there was a suture in situ on the left side of the neck at the junction of the internal jugular vein and the subclavian vein”* and *“there was no evidence of congestion or obstruction of the major blood vessels or the carotid arteries or jugular veins”*.
- The central venous cannula inserted into the right subclavian vein at induction for his transplant was situated in the right internal jugular vein and may have been obstructed, given unusually high CVP readings.
- Adam was described as ‘bloated’ when he left the operating theatre. It is uncertain whether this was generalised or confined to the head and neck. There are differing views on the presence of hypoxic damage or cellular reactive change in the brain.

The Inquiry’s Expert on Paediatric Anaesthesia, Dr. Simon Haynes has suggested that even in the absence of a suture, there may have been a degree of obstruction caused by the ‘scarring effect’ of a number of central venous lines having been inserted over time.

However, the Inquiry has been informed that the ligation of the left common facial vein did not involve the insertion of a suture in the left internal jugular vein, nor did the insertion of the Broviac line. Also, the Broviac line was removed in February 1995 and therefore considerably before Adam’s transplant surgery.

In addition, the Inquiry’s Expert on neuropathology, Dr. Wayne Squier has advised that any such constriction would have reduced overtime as other pathways would have been created.

- (i) What is your view on whether, given the information above, venous obstruction might have been present to a degree that it contributed significantly or at all to Adam’s cerebral oedema?
- (6) Describe and explain, to the best of your expertise, the likely progression or otherwise of cerebral oedema in a child who is being kept alive only by mechanical means. In particular:
- (i) Whether it was likely that Adam’s cerebral oedema would have progressed further after 11:55 on 27th November 1995 when he was reported as failing to awaken from anaesthesia and having fixed dilated pupils
 - (ii) If so the period over which cerebral oedema could have continue to progress and the further extent of cerebral oedema that would be likely over that period
 - (iii) Whether the cerebral oedema could have continued to progress right up until ventilation was abandoned 22 hours later

We refer you to Adam's history and early developmental record as set out in his medical notes and the records received from the South Eastern Education and Library Board ("SEELB"), in particular:

- 19th August 1991: Letter from Dr. Angela Bell to Dr. Maurice Savage which states that Adam's Apgar scores were 5 at 1 minute and 8 at 5 minutes. (Ref: 050-022-061)
- 28th November 1991: Radiological report states: "*Chest Normal heart shadow*" (Ref: 050-027-283)
- 8th December 1991: Following insertion of uteretic stents and a Broviac line and a laparoscopy, Adam is transferred from the theatre to PICU described as: "*pale and perfusion poor*" (Ref: 049-033-234)
- 22nd December 1991: Medical notes record: "*Phoned by nurse – Adam had an apnoeic episode ... Adam still not breathing when nurse went over to him – he was pale, required nurse to bang hard on cot side before respiration restarted*". See also: "*O/E Still pale. Breathing spontaneously ... in drawing + grunts, respiration irregular – pauses then rapid breaths*" (Ref: 050-023-068 and Ref: 050-023-069)
- 23rd December 1991: Medical notes record: "*9pm. Apnoea went off – breathing ceased but started with noise of cot side down. Dr. informed, morphine ↓1.5mls. Large green vomit immediately after. NG tube repositioned and left on free drainage. Hrly observations stable. Paracetamol given x2 for pain. Urine output good. Large ooze from around tubes*" (Ref: 051-023-128).
- 28th December 1991: Medical notes record: "*Mum noticed him twitching at 7.30pm. 'Jittery' – started in his legs, spread to involve his arms – sleeping at the time*" Similar entry in nursing notes. (Ref: 050-023-073, 051-023-132)
- 7th January 1992: Nursing notes record: "*Continues to vomit overnight. Pyrexia this am. One episode of being "jittery" during the night*" (051-023-136).
- 7th February 1992: Ultra sound following which the radiological report states: "*The heart is enlarged. Pulmonary vascularity is slightly increased*" (Ref: 050-031-290)
- 7th February 1992: Medical notes record: "*Problems ? Facial oedema*" (Ref: 050-023-087)
- 8th February 1992: Medical notes record: "*Mum said Adam had transient twitching of L eye. Went pale. No tonic clonic movement of limbs*" (Ref: 050-023-088). Nursing notes record: "*9.2.92 7am ...? twitching episode last pm. Mum watching him, said his head jerked and left eye flickered for few secs. Colour appeared pale. BM checked 4mmols. Seen by Dr. Sanhi to observe closely for any further*

- episode otherwise settled night*" (Ref: 051-023-146 to 051-023-147)
- 4th March 1992: X-ray report following which the radiological report states: "*CHEST The heart is slightly enlarged. Pulmonary vascularity appears normal. The lungs are clear*" (Ref: 052-026-169)
 - 9th March 1992: X-ray report following which the radiological report states: "*CHEST The heart is a good deal smaller. Pulmonary vascularity appears normal. Lungs clear*" (Ref: 052-026-168)
 - 10th April 1992: X-ray report following which the radiological report states: "*CHEST The heart has increased in size. The lungs are clear*" (Ref: 053-033-148)
 - 14th April 1992: X-ray report following which the radiological report states: "*Chest The heart is slightly smaller. The lungs appear clear*" (Ref: 053-033-147)
 - 13th May 1992: X-ray report following which the radiological report states: "*CHEST The heart remains slightly enlarged. Pulmonary vascularity is normal. Lungs clear*" (Ref: 053-029-141)
 - 21st May 1992: X-ray following which the radiological report states: "*The heart remains slightly enlarged. The lungs are clear*" (Ref: 055-055-240)
 - 23rd October 1992: X-ray following which the radiological report states: "*Knees and Wrists. There is some generalised osteopaenia but no definite change of renal osteodyseryrophy*" (Ref: 055-055-238)
 - 19th December 1992: Medical notes record: "*Attended ward. 2.30pm. Mum concerned re puffiness and vomiting. Woke this am side of face puffy ... doesnt appear puffy*" (Ref: 054-057-150)
 - 4th March 1993: Medical notes record: "*CXR – lung fields clear. Rotated ? heart enlarged*" (Ref: 052-023-047)
 - 5th May 1993: X-ray report following which the radiological report states: "*Chest The heart size is within normal limits. Pulmonary vascularity is normal. The lung fields are clear*" (Ref: 057-113-331)
 - 14th December 1993: X-ray following which the radiological report states: "*Hands, wrists and knees There is general reduction in bone density with cortical thinning and there appears to be early erosive changes in the distal ulnar metaphyses suggesting early renal osteodystrophy*" (Ref: 057-113-330)
 - 5th September 1994: Medical notes record: "*C/O sore head this pm. "hearing noises in ears".*" (Ref: 057-102-184)

- 18th January 1995: Bacteriology report states: *“clinical diagnosis and reason for request: pyrexia rigors, peritoneal dialysis”* (Ref: 057-105-264)
- 2nd February 1995: Medical notes record: *“No rigors or shakey episodes”* (Ref: 057-102-188)
- 29th June 1995 and 1st July 1995: X-ray report following which the radiological report states: *“Heart size and pulmonary vascularity normal. The lungs are clear”* (Ref: 057-108-324)
- 17th August 1995: X-ray report following which the radiological report states: *“Pelvis Early changes of renal osteodystrophy. No other bony abnormality”* (Ref: 058-048-245)
- 9th November 1995: X-ray report following which the radiological report states: *“Left hand and wrist No evidence of renal osteodystrophy. Bone age is 2 ½ years”* (Ref: 058-046-243)

(i) Is there anything in those records that could have affected the development of dilutional hyponatraemia in Adam?

(7) What effect (if any) could any impairment to Adam’s cerebral blood flow, whilst he was in the operating theatre, have had to the level of his cerebral oedema when he left there? If it could have had an effect on it, then what contribution (if any) could that have made to his ultimate gross cerebral oedema, assuming that he was adequately oxygenated in PICU?

(8) Your attention is also drawn to the following:

- Dr. Wayney Squier says that she did not regard the cerebral oedema as the most extreme that she had seen. However, she cautioned that the photographs were taken 24 hours after the surgery and that the further activity may have reduced the oedema. She sought input from a Consultant Radiologist Dr. Philip Anslow who examined a CT scan of Adam’s brain taken in July 1995 and another taken 2 hours after the surgery. He concluded that the changes he saw in the CT scan taken after surgery were particularly severe in the posterior fossa but that he believed the appearances were in keeping with the development of brain oedema and were sufficient to have been fatal.
- Chest x-rays taken at 13:20 and 21:30 after the transplant surgery were thought by the clinicians to show pulmonary oedema. This was initially queried by the Inquiry’s Expert Consultant Radiologist, Dr. Caren Landes but following the receipt of clearer images she advised that the x-

ray taken at 21.30 did show evidence of pulmonary oedema (Inquiry reference x)[*please insert*]²⁵

- Inquiry Witness Statements from Dr. Les Dyer, a retired Anesthesiologist who assisted Adam's family in understanding technical matters after the Inquiry had been established
- An Inquiry Witness Statement from Dr. Robert Taylor who was the Consultant Paediatric Anaesthetist involved in Adam's transplant surgery, in which he changed his position
- The reports of the Inquiry's Experts Professor Peter Gross, Dr. Malcolm Coulthard, Dr. Simon Haynes, Professor John Forsythe & Mr. Keith Rigg who all deal with aspects of fluid management or issues that affect fluid management
- Comparative tables prepared by Professor Peter Gross and Drs. Malcolm Coulthard, Simon Haynes and Robert Taylor of the fluid calculations that were made for Adam's transplant surgery

In the light of all the material that has been provided to you and all the issues to which your attention has been drawn, please state and fully explain your opinion as to whether Adam's Cerebral Oedema and subsequent death was caused:

- (i) Solely by hyponatraemia
- (ii) Mainly by hyponatraemia with other contributory causes, and if with other contributory causes please detail them and explain their significance
- (iii) By a number of conditions (ie multifactorial causation) of which hyponatraemia was a material contribution, and if so detail the conditions and explain their significance

Conclusion

40. It is of fundamental importance that the Inquiry receives a clear reasoned opinion on these issues.
41. Your assistance on the Inquiry's requirements should be provided in the form of a fully referenced Expert's Report.

²⁵ The Inquiry has also received a Report from a Dr. Louise Sweeney, also a Consultant Paediatric Radiologist, which states in respect of the x-ray at 13:20: "*mild pulmonary oedema mainly in the right perihilar region*" and in relation to the x-ray at 21:30: "*increase in heart size and a deterioration in the appearance of the lungs due to an increase in pulmonary oedema in both lungs with evidence of interstitial and intra-aveolar pulmonary oedema*". Ref: WS-242-1, p.2

INDEX OF KEY ACCOMPANYING DOCUMENTS

Tab.1 Brief

Tab.2 Selected Inquest Documents:

Depositions:

- Dr. Maurice Savage (Ref: 011-015-109)
- Dr. Alison Armour (Ref: 011-010-030)
- Dr. John Alexander (Ref: 011-012-079)
- Dr. Robert Taylor (Ref: 011-014-096)

Reports:

- Dr. Alison Armopur's Report on Autopsy (Ref: 011-010-034)
- Professor Peter Berry (Ref: 011-007-20)
- Dr. Edward Sumner (Ref: 011-011-042)
- Dr. John Alexander (Ref: 011-012-084)

Letters/Notes:

- Coroner's note on Adam Strain case dated 8th December 1995 (Ref: 011-025-125)
- Letter from Dr. Alison Armour to Professor Berry requesting a report on Adam Strain's case for the Coroner (Ref: 011-029-151)

Tab.3 Selected PSNI Documents:

Statements:

- Ms. Debra Slavin (Ref: 093-005-007) and attached photographs of Adam (Ref: 093-005-108 to Ref: 093-005-113)

Reports:

- Professor Peter Berry (Ref: 093-030)
- Professor Risdon (Ref: 093-031)
- Medical opinion of Dr. Edward Sumner, including PSNI brief (Ref: 094-001-001 and Ref: 094-002-002)

Other documents:

- Dr. Alison Armour's Notification of Autopsy findings (Ref: 094-114-321)

Tab.4 Selected Inquiry Documents & Materials:

Initial Witness Statements of:

- Ms. Debra Slavin (1st)
- Dr. Maurice Savage (1st, 2nd, 3rd & 4th)
- Mr. Patrick Keane (1st, 2nd & 3rd)
- Dr. Robert Taylor (1st, 2nd, 3rd & 4th)
- Dr. Mary O'Connor (1st, 2nd, 3rd & 4th)
- Dr. Edward Sumner (1st)

Other Inquiry Documents & Materials:

- Letter dated 2nd March 1995 from Dr. Maurice Savage to Adam's GP, Dr. Scott (Ref: 057-072-133)
- Laboratory report of serum sodium result of 133mmol/l from blood taken sometime on 26th November 1995 (Ref: INQ-0450-11)
- Laboratory report of serum sodium result of 133mmol/l from blood taken sometime on 26th November 1995 (Ref: INQ-0450-11)
- Autopsy Request Form
- Letter dated 10th May 2011 from State Pathologist's Office to the Inquiry attaching the contemporaneous notes of Dr. Alison Armour
- Adam's medical notes and records for 26th November 1995 - 29th November 1995
- Armour - Dilutional hyponatraemia: a cause of massive fatal intraoperative cerebral oedema in a child undergoing renal transplantation, *J Clin Pathol* 1997; 50:444-446
- CT-scan of Adam's brain taken after his transplant surgery on 27th November 1995
- Photographs of Adam's brain following fixing that are included in Dr. Alison Armour's article (Figures 1 & 2)
- 2 post-transplant surgery chest x-rays