CORONERS COURT
OF NEW SOUTH WALES

Inquest:	Inquest into the death of Ezekiel Howard
Hearing dates:	24-28 October 2016
Date of findings:	8 December 2016
Place of findings:	State Coroner's Court, Glebe
Findings of:	**Deputy State Coroner, Magistrate Teresa O'Sullivan**
Catchwords:	CORONIAL LAW – Cause and manner of death Sudden Unexpected Death in Epilepsy (SUDEP) Sudden Cardiac Death (SCD) Sudden unexpected death of a child Back-up of Medical records *Sudden Death in Epilepsy – Forensic and Clinical Issues* – Claire M Lathers and Ors CRC Press *2011* Chap 7,19,21, 24 Tomson et al, Sudden Unexpected Death in Epilepsy: A Review of Incidence and Risk Factors, *Epilepsia* 46 (Suppl.11) 45-61, 2005 Nouri, et al, Sudden Unexpected Death in Epilepsy: Overview, Autopsy Findings, E-medicine, *Medscape*, December 2015 Bagnall, Semsarian and others, New England Journal of Medicine – A Prospective Study of Sudden Cardiac Death in Children and Young Adults – *N Engl J Med* 2016; 374:2441-52 Bagnall, Semsarian and others, Research article: *Annals of* *Neurology* – Exome Based Analysis of Cardiac Arrhythmia, Respiratory Control and Epilepsy Genes in Sudden Unexpected Death in Epilepsy – 2016 American Neuological Association

File number:	2011/390602
Representation:	**Ms Kirsten Edwards, Counsel Assisting, instructed by Ms Janet de Castro Lopo, Office of General Counsel** **Mr Graham Segal for the Howard Family** **Mr Richard Sergi for the Nepean Blue Mountains LHD and Mr Hinrichsen** **Ms Eva Elbourne for Dr Kolos** **Mr Todd Pickering for Dr Murphy**
Findings:	**Identity of deceased**: The deceased person was Ezekiel Howard. **Date of death**: He died on 12 September 2011. **Place of death**: He died at Colo Heights in NSW. **Manner and cause of death:** The death was sudden and unexpected. The evidence does not allow me to identify the cause of death.

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The Coroners Act 2009 (NSW) *in s81 (1) requires that when an inquest is held, the coroner must record in writing his or her findings as to various aspects of the death.*

These are the findings of an inquest into the death of Ezekiel Howard.

Introduction:

1. Ezekiel (Zeke) Howard, a ten year old boy, died in his sleep on 12 September 2011. Zeke's mother, Mrs Sonja Howard, found Zeke unresponsive in his bed in the morning when she went to wake him for school. It was about 7:30am. Mrs Howard said Zeke was in bed on the bottom bed of a bunk bed. The bed clothes were not disturbed and he seemed to be in his normal sleep position. Zeke was stiff and cold to touch. There was vomit around his pillow. Mrs Howard called for her husband, William, and put Zeke in a warm shower to try and revive him. An ambulance was called and paramedics attended but there was no attempt at resuscitation.

2. At the time of his death Zeke was a reasonably healthy child. He had asthma but no other diagnosed conditions or illnesses.

3. He spent the day before his death riding his trail bike around his family property at Colo Heights and there is no suggestion of any accidents or falls. He had a normal dinner and watched TV with his family and didn't complain of feeling unwell or look sick. He hadn't been ill that week.

4. His death was sudden, unexpected and incredibly tragic. It has devastated his mother, Sonja, his father, William, and his little sister Epiphony.

5. The circumstances of his sudden and unexpected death have raised many questions for his family about what the underlying cause of his death was. This inquest has been largely concerned with the manner and cause of Zeke's death. I hope some of the family's questions have been answered. I thank them for their participation in this emotional and lengthy process.

The Inquest:

6. An inquest is different to other types of court hearings. It is neither criminal nor civil in nature and the coroner does not make determinations and orders that are binding on the parties, such as in civil litigation, nor determine whether a person is guilty or not guilty of an offence, such as in criminal proceedings.

7. The formal findings that need to be made are: who, when, where someone died and the manner and cause of their death. It is not always possible to definitively answer each of those questions.

8. We know the answer to the first three questions. Zeke Howard died at home at Colo Heights, New South Wales, on 12 September 2011. The focus in this inquest was on the manner and cause of Zeke's death. Zeke's sudden and unexpected death raises questions about both the immediate mechanism of death and its underlying cause. Both were described as "unascertained" at autopsy. As I will outline below, most of the evidence at the inquest related to whether Zeke could have died from Sudden Death in Epilepsy (SUDEP) or Sudden Cardiac Death (SCD).

Zeke's Medical History:

Early history

9. Zeke was born on 22 August 2001 at Hawkesbury Hospital. He had a vacuum assisted birth. Zeke experienced prolonged tachycardia (increased heart rate) during Mrs Howard's labour and Mrs Howard had to alert staff to her concerns that her labour was abnormal. After first being reassured by nurses that it was normal to feel anxious during labour, it was discovered that Zeke was indeed in trouble and there was a need for medical intervention. As Mrs Howard told me during her family statement, this experience was the first of many in Zeke's tragically short life where Mrs Howard felt that her concerns about his health were dismissed or minimized by the medical profession only for subsequent events to prove her right. I am very sympathetic to Mrs Howard's anger and distress at not being carefully listened to when it came to the health of her child.

10. When Zeke was 3 months old he was admitted to Hawkesbury Hospital (on 23 November 2001). Zeke's GP Dr Rex Stubbs (now deceased) sent a letter to the Hospital stating that Zeke had experienced a few minutes of being unresponsive. Medical notes stated that Mrs Howard could not rouse Zeke by shouting at him and Zeke had a large vomit when he was roused. The letter also stated "there was a similar episode at the age of 3 weeks". A brain ultrasound conducted on Zeke on 26 November 2001 was reported as normal. No focal lesions or congenital abnormality was detected. Epilepsy was one possibility explored in differential diagnosis. An EEG and sleep studies were considered. Mrs Howard said she was not aware of any EEG test being arranged or a follow up appointment. Mrs Howard ultimately discharged Zeke against medical advice. Mrs Howard said she did so after being quizzed aggressively about whether or not she had shaken Zeke. Again Mrs Howard had an unpleasant experience in the medical system at a time when she was highly anxious about the health of her child.

11. Zeke had a number of other visits, eight in total, to Hawkesbury Hospital ED in his first two years. These attendances mostly related to asthma and bronchial problems. I mention them not because they are likely to be related to the cause of death (no expert suggested Zeke's asthma was related to the cause of death) but because they demonstrate that Mr and Mrs Howard were diligent parents who sought help pro-actively when they were concerned about their child. It appears

from the notes and Mrs Howard's evidence that some of the experiences they had in Hospital during this time were stressful and unpleasant.

12. Zeke's health appeared to improve after he was a toddler although the family moved to England for a time to assist with his asthma. The Howard family returned in time for Epiphony's birth in early 2006.

Events in 2011

13. In April and May 2011 (when Zeke was 10) Zeke had some episodes which could have been related to his death. On 21 April 2011 Zeke experienced an episode of left side weakness, facial droop and slurred speech. Mrs Howard took Zeke to GP Dr David Morris. Mrs Howard said that the symptoms resolved after a few minutes. Dr Morris ordered some blood tests and recommended Mrs Howard return after the results were received. It appears the blood test results were normal. Zeke was admitted to hospital before the family could return to see Dr Morris.

14. On 6 May 2011 Mrs Howard heard a bang and rushed to Zeke's bedroom to find him crying on the floor. He was unable to pull himself up. He had wet himself and there was blood in the side of his mouth. Zeke said he had fallen while climbing down the top bunk to go to the toilet. Mrs Howard rushed Zeke to hospital. On the way she noticed that Zeke had facial droop on the left side of his face and slurred speech. Mrs Howard had recently been trained in the symptoms of stroke and was very concerned Zeke was experiencing a stroke. After Zeke and Mrs Howard arrived at Hawkesbury Hospital Zeke vomited 3 times. Mrs Howard said they spent an extremely long time waiting first at triage and then to be seen by a doctor at the Hospital. The wait must have been excruciating, particularly in light of Zeke's history and Mrs Howard's conviction that he was suffering from a life-threatening event.

15. Zeke was admitted under the care of consultant paediatrician Dr Phillip Kolos. He was seen by a number of doctors including Dr Kolos, Dr Birgit Tauber and paediatric registrar Dr Kevin London. The facial droop and slurred speech had resolved by the time Zeke was examined but he complained of a mild right-sided headache. There was obvious limb weakness and he initially needed assistance to walk.

16. The medical notes record a history of similar events. One note states: "Mum states similar episode a few weeks ago, similar symptoms but resolved after a few minutes" (I assume this note refers to the episode on 21 April 2011). The notes also record:
    - "*check w GP 14/7 ago because of similar events, 3-4 events year for some years, always short 1-2 min and self-resolving, never incontinent, [never?] tongue bite, mother did not follow up*"
    - "*Patient has been having 3-4 episode in the last 2-3 years, unknown to parents*"

- "*1/12 noticed facial droop for few minutes, not associated w headache not usually having headache, mother gets facial droop from time to time…mo side – epilepsy – not (?) of weakness*"

17. Mrs Howard told the inquest that she was not aware of the episodes until she heard Zeke tell the doctors about them. She said she heard Zeke say the episodes involved tingling in his hands and feet. She said her facial droop related to a facial muscle injury from her birth and could not be connected to epilepsy. She said she told the doctors she had a family history of epilepsy but later discovered that history was not accurate.

18. Dr Kolos recommended the following investigations which were recorded in a discharge plan on 7 May 2011:
    1. MRI/MRA at Nepean Hospital
    2. EEG at Westmead Children's Hospital
    3. Blood tests at Nepean Hospital
    4. Cardiology clinic at Nepean Hospital
    5. See Dr Kolos

19. The experts at the inquest agreed that this diagnostic investigation pathway was appropriate.

Investigations in 2011

20. Zeke had an electro cardiogram (ECG) examination of his heart on 6 May 2011. The ECG computerised report read as follows:

    "*Normal sinus rhythm with marked sinus arrhythmia. RSR pattern illegible suggests right ventricular conduction delay". Borderline ECG"*

21. The ECG test displayed a normal corrected QT interval and no ECG patterns for Brugada syndrome. The reference to right ventricular delay suggested that Zeke could suffer from "incomplete right bundle branch block". As I discuss below, Professor Thomas was of the opinion this finding could be related to Zeke's cause of death. The cardiologist experts gave evidence at the inquest that the finding was very common in children. Dr Kolos reported that the ECG was normal for Zeke's age. The expert cardiologists at the inquest agreed that the results were normal for Zeke's age.

22. I note here that the expert evidence was to the effect that this result did not exclude any heart condition, including conditions known to cause sudden death like Long QT syndrome and Brugada syndrome. The test simply did not detect any condition.

23. Zeke also had a CT brain scan 6 May 2011. The findings were reported as normal. On 18 May 2011 Zeke had the MRI of his brain and a MR angiogram. It

was reported as normal with "*no intracranial mass lesion or MR features to suggest vascular malformation".* This finding is significant because, in conjunction with the autopsy results, it suggests that Zeke's death was not connected to the hole in his heart (this is discussed further below).

24. On 25 May 2011 Zeke saw paediatric cardiologist Dr David Murphy. Dr Kolos sent Dr Murphy a referral letter which noted Zeke had presented with multiple focal neurological symptoms. Dr Kolos asked Dr Murphy to perform an echocardiogram (a heart ultrasound) to screen for a heart defect. Dr Murphy was informed that Zeke had a family member with a hole in the heart.

25. In Dr Murphy's reporting letter to Dr Kolos, signed the same day, he recorded:

    "…*Echocardiography today demonstrated normal cardiac structure and function. His atrial and ventricular septum were intact and there was no obstruction to intracardiac flows. His aortic valve was trileaflet and gave rise to a normally branching left sided aortic arch. His systemic venous return superiorly was normal and unobstructed. His myocardium inherently appeared normal and there was no pericardial effusion. He had good systolic function with an LVED of 45mm, fractional shortening of 45% and free wall 7mm.*

    The findings today pleasingly confirm that Ezekiel's heart is structurally and functionally normal, with no cardiac aetiology contributing to his neurological symptoms and left partial hemiparesis…."

26. It appears that Dr Murphy took a personal call during the consultation with Zeke. He also expressed the view that Zeke should lose some weight. These comments were subject to a complaint to the HCCC. Dr Murphy did not give evidence to the inquest and was not requested for cross-examination by the Howard family. His response to the HCCC implicitly accepts a personal call was taken during the consultation.

27. I make the following observations about the consultation with Dr Murphy.

28. First, the echocardiogram (echo) did not detect a hole in the heart. It was discovered on autopsy that Zeke did in fact have a small hole in the heart known as a Patent Foreman Ovale (PFO). The experts did not consider the PFO had any relationship to the cause of death, nor were they critical of the failure to detect the small hole during the echo test.

29. Secondly, the letter records a measurement for the left ventricular free wall of 7mm. The left ventricular free wall was measured at 14 mm at autopsy. The expert evidence was to the effect that the discrepancy was to be expected as post-mortem increases in size are normal. The thickness was described as "on the high side of normal" for Zeke's age group.

30. Next, the letter was extremely reassuring to Mr and Mrs Howard. Zeke's family understandably viewed the letter as a clean bill of cardiac health. They viewed the letter as excluding the possibility that Zeke suffered from any serious cardiac problem, let alone that he was at risk of sudden death in the next few months.

31. Zeke's death would have been confronting in any circumstances but, as Mr Segal, counsel for the Howard family eloquently expresses in his submissions, the family's sense of reassurance was shattered by Zeke's death. Zeke's unexpected death has caused them to reflect considerably upon their consultations with Dr Kolos and Dr Murphy. Their faith and trust in the medical system has been irrevocably destroyed.

32. I agree with Dr Murphy's counsel that there is no reason to believe that Dr Murphy's measurements were inaccurate. I equally have no reason to conclude that the content of his report was anything but accurate and appropriate. There is no evidence before me which would allow a conclusion that Zeke died from a heart condition which should have been detected before his death.

33. However, I also completely understand why the Howard family have agonised over the failure to detect the PFO and queried if the left ventricular measurements were correct, or perhaps an error caused by distraction due to the interruption of the personal call. I believe many in their circumstances would feel angry and betrayed and raise similar questions.

34. As I will detail later in this finding, the final blow to whatever was left of the Howard family's faith in the system came when it was discovered that original image of the echocardiogram was inadvertently deleted due to a highly unsatisfactory failure by Nepean Hospital to properly store images recorded on the ultrasound machine.

35. For completeness, I should note that Zeke never underwent an EEG examination. It is very clear that Mrs Howard has suffered from exceptional distress because she has contemplated that, if Zeke's death was connected to undiagnosed epilepsy, she might somehow be responsible due to her failure to attend the test. Mrs Howard has detailed her reasons why she did not attend the test with Zeke and they are understandable. Epilepsy expert Dr Lawson was also critical of the many structural impediments to families, particularly those in the outer suburbs, accessing EEG tests in NSW. I have not made any recommendations about EEG tests in this inquest for procedural fairness reasons but it may be the subject of further investigation in this Court. In any event, I understand from Mrs Howard's counsel's submissions that the evidence at the inquest has put her mind to rest in this regard. I sincerely hope that is the case.

Investigations After Death:

36. A post mortem examination on Zeke was conducted on 13 September 2013. The autopsy was conducted by Dr Kendall Bailey and supervised by Professor Jo Du

Flou. Specialist neuropathologist Dr Michael Rodriguez dissected and analysed the brain. Blood, bacterial, virology and toxicology tests were also conducted. The post mortem report concluded that the death was “unascertained”.

37. In 2013 a post-mortem blood sample containing Zeke’s DNA was sent to Harvard Partners in Boston and Oslo University genetic testing centre in Norway. A total of 59 cardiac genes were tested. The genes were those related to cardio-myopathies, primary arrhythmogenic disorders such as long QT syndrome, Brugada syndrome and CPVT, as well as three genes related to congenital septal defects.

38. The testing did not identify any clinically significant DNA variants. The testing did not reveal any gene mutations responsible for the common causative genes in long QT syndrome, Brugada syndrome or CPVT. One DNA variant was identified in the CSRP3 gene (Trp4Arg). In his original report in 2013 Professor Semsarian noted that the variant was believed to be associated with cardio-myopathy. However, he informed the inquest that subsequent international research has concluded the gene variant is benign.

The Experts:

39. The inquest has had the benefit of seven expert reports. One pathologist, Professor Thomas, gave oral evidence by telephone and three experts, Doctors Roberts and Lawson and Professor Semsarian gave simultaneous evidence in a conclave.

40. The reports and oral evidence explored:

a) the immediate circumstances of Zeke’s tragic death;
b) Zeke’s demographic and risk factors for sudden death;
c) Zeke’s medical history;
d) the results of investigative tests, particularly those conducted in 2011 including a brain MRI, CT scan, ECG and echocardiogram;
e) the pathological findings at autopsy;
f) the results of brain dissection by a specialized neuro-pathologist;
g) Zeke’s family history; and
h) the results of genetic testing.

The conclusion of the experts in their reports

41. **Dr Kendall Bailey** is a forensic pathologist employed by the Department of Forensic Medicine. Dr Bailey conducted the post-mortem examination on Zeke on 13 September 2011. She determined that:

"[*B]ased on the autopsy findings, including evaluation of the toxicology, microbiology and biochemistry reports, and review of the medical records, the cause of death remains unascertained. The medical history, neuropathologist's findings and absence of other significant pathology are suggestive of epilepsy and it is likely that this is the underlying cause of death in this case. However while SUDEP (Sudden Death in Epilepsy) is a well recognized phenomenon, statistically affecting young males with poorly controlled epilepsy, in the absence of a clear clinical diagnosis, epilepsy should not be given as a cause of death in this case…While SUDEP is a likely cause of death in this case, an underlying cardiac condition cannot be excluded. Cardiac death may occur suddenly with little or no morphological changes, resulting in negative autopsy findings."*

42. Dr Michael Rodriguez is a specialist neuropathologist also employed by the Department of Forensic Medicine. He provided a report of his examination of Zeke's brain on 11 January 2012. That report included the comment:

    "[*A]lthough mild and non-specific, many of the lesions identified in this case may be seen in patients with epilepsy. However, several, including the slight hippocampal asymmetry, and microdysgenesis in the amygdala, have also been described as incidental findings in otherwise normal brains."*

43. **Acting Professor Michael Cheung** is the Director Department of Cardiology, Royal Children's Hospital Melbourne. Professor Cheung provided a letter to Detective Atkins, the police officer in charge of the coronial investigation on 21 August 2011. The letter did not express an opinion on the cause of death but concluded that there was no evidence of negligence in Zeke's case.

44. **Dr Phillip Roberts** is a fulltime paediatric cardiologist employed at the Heart Centre for Children at the Children's Hospital at Westmead. He was retained directly by the Coroner's Court. In his report dated 20 April 2012 he concluded:

    "From the information and records provided I would conclude that Ezekiel Howard had suffered a neurological event for which a list of differential diagnoses had been established and an appropriate investigative pathway embarked upon. Sadly his death did not allow for the completion of the tests arranged…I concur with the autopsy report that SUDEP and an arrthymic cardiac death cannot be excluded as mechanisms for death and one may lead to the other particularly if there was already evidence of some inflammatory process in the heart".

45. Professor Johan Du Flou is a forensic pathologist who at the time of his report was Chief Forensic Pathologist at the Department of Forensic Medicine and a Clinical Professor at Sydney University. He has also co-authored research studies on Sudden Cardiac Death and Sudden Death in Epilepsy. Professor Du Flou was retained by the Coroner's Court. In his report dated 4 September 2012 he concluded:

    "[I]n summary, I do not believe that there are any major differences in opinion between the various medical experts [Bailey, Cheung, Roberts]

consulted in this case. The cause of death remains unexplained but is a sudden death which is likely either the consequence of a neurological abnormality such as epilepsy or a primary cardiac rhythm abnormality, or a combination of or interaction between the two."

46. **Professor Christopher Semsarian** is an adult cardiologist with an internationally renowned expertise in the genetic investigation of sudden and unexpected death in children and young adults. He was the leading author of the two largest Australian research studies on Sudden Cardiac Death and Sudden Death in Epilepsy. He was retained directly by the Howard family. Professor Semsarian stated in a letter dated 27 June 2013:

    "[I]n my opinion, based on the collective information from the clinical history, the post-mortem findings, and the genetic studies, there is no definitive cause of death in Ezekiel Howard. Based on our current knowledge of sudden death in the young, an underlying cardiac disorder is a major cause, of which the majority are inherited (genetic) heart diseases…I note the possibility of SUDEP has been raised as a possible cause of death…In the absence of a definite clinical diagnosis of epilepsy in Ezekiel during life, I think SUDEP would be a very unlikely cause of death"

47. **Professor Tony Thomas** is an anatomical pathologist and Senior Specialist and Head of Department in Anatomical Pathology at Flinders Medical Centre. He is a Professor of Pathology at Flinders University. He has a particular interest in cardiac pathology. He was retained directly by the Howard family. In his report dated 14 March 2014 he concluded:

    [B]ased on the post-mortem macroscopic and microscopic findings provided, the cause of death is not obvious and is best considered as unascertained. In that there were subtle cardiac and neuropathological changes, it is appropriate to consider cardiac and neuropathological causes of sudden unexpected death…[notes the subtle abnormal inflammatory changes in heart, the non-specific changes in the brain, the absence of any gene mutation associated with epilepsy and a gene variant associated with cardiomyopathy] *In my opinion, it therefore seems more likely that Ezekiel suffered an arrhythmic cardiac death related to an underlying cardiomyopathy than sudden unexpected death due to epilepsy*"

48. **Dr John Lawson** is a specialist paediatrician and neurologist and full-time staff specialist in paediatric neurology at Sydney Children's Hospital, Randwick. He has particular expertise in childhood epilepsy. He was retained by the legal team assisting the Coroner in the inquest. Dr Lawson's report dated 5 December 2015 stated:

    "I do not believe there is sufficient information to definitely diagnose the cause of Ezekiel's death. It is my opinion that the child very likely had untreated focal epilepsy. The main assumption on which this is based is the limited history of events provided in the documents by the mother and the hospital records, which are largely concordant. This clearly is different

to the usual practice of diagnosing epilepsy which is a direct and timely discussion with the family, child and any other witnesses. This means that in the absence of another clear explanation then SUDEP is a likely cause of death. This would meet the criteria below except that the child did not have a recognised diagnosis of epilepsy during life."

49. It must be noted that the reports were obtained over the course of four years and were not compiled with reference to the same information. Some relevant information changed after reports were completed. Dr Bailey, Dr Roberts and Professor Du Flou all provided reports on the assumption that Zeke had a maternal history of epilepsy. Zeke's mother, Mrs Howard later provided a statement to the effect that she did not believe that history had been accurate.

50. Professors Semsarian and Thomas wrote their reports on the (superseded) assumption that the gene variant Trp4Arg was associated with cardio-myopathy. Further, Professors Semsarian and Thomas did not have access to the medical notes relating to Zeke's episodes as a baby or in 2011. They were not aware at the time of their reports that Zeke had complained of 3-4 "episodes" a year for several years. Professor Thomas also believed that genetic testing had excluded epilepsy.

51. The experts had access to the entire brief of evidence before they gave oral evidence. In oral evidence Professor Thomas, Professor Semsarian and Dr Roberts all stated that their ultimate opinion was not altered by the new information but had some ameliorating effect on whether they considered epilepsy or cardiac causes more likely, that is made the cause of death more likely to be "undetermined".

The evidence relating to Sudden Unexpected Death in Epilepsy (SUDEP):

52. SUDEP is not a cause of death but a label to describe sudden and unexpected deaths which occur in people who have been diagnosed with epilepsy. The definition of a SUDEP death is:
    - the victim had epilepsy, defined as recurrent (defined as more than one) unprovoked seizures;
    - the victim died unexpectedly while in a reasonable state of health;
    - the death occurred "suddenly" (in minutes), when known;
    - the death occurred during normal activities (e.g., in or around bed, at home, at work) and benign circumstances; and
    - an obvious medical cause of death was not found.

53. Zeke's death could never technically be described as SUDEP because he did not receive a clinical diagnosis of epilepsy during life. The inquest explored the possibility that Zeke had undiagnosed epilepsy which placed him at risk of sudden death and may have caused his death.

54. People diagnosed with epilepsy, including children, are known to be at greater risk of sudden death than the general population.[1] The mechanism of death in cases of SUDEP is still being explored and the mechanism may not always be the same. There is evidence that at least some of the people who die of SUDEP have an underlying cardiac condition. Professor Semsarian's studies found a number of people who died of SUDEP had long-QT syndrome.[2] The genetic connection between SUDEP and other cardiac conditions is also still being explored.

55. There is evidence that people with an underlying cardiac condition can be misdiagnosed with epilepsy because cardiac conditions can manifest with symptoms which are mistaken for epileptic symptoms.

56. Dr Lawson informed the Court that epilepsy is a brain disorder of episodic electrical events that result in a clinical change. There are thousands of different types of epilepsy. Not all epilepsy involves the type of tonic clonic seizures usually associated with epilepsy in popular culture. Epilepsy cannot be definitively diagnosed by a test. Dr Lawson stated that the best method to diagnose epilepsy is from a clinical history. The diagnosis can be supported by investigative tests which include MRIs and EEGs, neither of which are considered definitive. EEGs have only a 50% chance of detecting epilepsy because the test relies on an occurrence of abnormal brain activity as the test is being conducted. MRIs do not detect epilepsy but can detect brain abnormalities which cause epilepsy.

57. Genetic testing and family history can also inform an epilepsy diagnosis but Dr Lawson said they do not play a significant role. A family history of epilepsy might be supportive of a diagnosis but Dr Lawson stated that many people with genetic epilepsy do not have a family history of epilepsy. There are many genes associated with epilepsy and not all can be identified through genetic testing. Genetic testing of Zeke's DNA was negative for one genetic component (SCNA1) known to be associated with epilepsy and SUDEP. That testing does not preclude Zeke from having epilepsy or from having other genes associated with epilepsy. Professor Semsarian and Dr Lawson both referred to 72 genes known to be linked to epilepsy and SUDEP.

58. Dr Lawson stated that he believed that there was a very strong chance that Zeke had undiagnosed focal epilepsy. Dr Lawson based this assessment on the medical notes relating to the two occasions when Zeke was not rouseable as a baby, his two experiences with transient left hemiparesis (short-lived loss of functionality of his left side) in 2011 and the medical notes of Zeke suffering from

[1] In children diagnosed with epilepsy who were followed for 40 years, 25% died with 38% of deaths ascribed to SUDEP, Semsarian et al "Exome-Based Analysis of Cardiac Arrhythmia, Respiratory Control, and Epilepsy Genes in Sudden Unexpected death in Epilepsy, 2016 American Neurological Association, p2

[2] Long-QT syndrome is a genetic heart condition known to cause sudden death. Genetic and clinical testing did not find any evidence Zeke had long-QT syndrome but those tests cannot definitively exclude the condition.

3-4 episodes a year for “several years”. Weakness on one side of the body is a known symptom of focal epilepsy. Cardiac conditions which mimic epilepsy tend to cause brief periods of unconsciousness or “syncope” rather than focalized weakness. (But the experts agreed that a cardiac cause for the events could not be excluded.)

59. Dr Lawson readily volunteered that his confidence that Zeke had undiagnosed focal epilepsy (expressed in his report as a 95% chance) was based on incomplete and imperfect information. There was very little information about the “3-4 episodes for several years”. The notes do not make clear what features of the episodes were “similar” to the events on 21 April and 6 May 2011. Dr Lawson said the tingling Mrs Howard said Zeke described might be indicative of epilepsy and that focalized tingling on one side would be of particular significance. Dr Lawson said he would normally defer diagnosis until he received a detailed patient and family history including witness accounts or ideally video footage of seizure events. In essence, Dr Lawson was really saying that if Zeke had presented to him as a patient he would have strongly favoured a diagnosis of epilepsy in a process of differential diagnosis and conducted further investigation. Zeke’s tragic death prevented further investigation.

60. If Zeke had epilepsy he was more at risk of sudden death, but it does not follow that Zeke’s death was caused by epilepsy even if he had the condition. Dr Lawson said in the conclave that he believed that the cause of death was undetermined and there was not enough information to assign a cause of death. He agreed that death from epilepsy was a very rare occurrence in children of Zeke’s age.[3] Zeke had few of the risk factors associated with SUDEP. In particular, SUDEP occurs more often to children who suffer from neurological deficits and those who suffer from chronic tonic clonic seizures. Dr Lawson said a death from epilepsy in the combined circumstances of Zeke’s clinical history and age was “extraordinarily rare” but could not be discounted. He also did not discount the possibility of a cardiac condition.

61. Professor Semsarian and Dr Roberts agreed with Dr Lawson. Dr Roberts said in the conclave that he would not exclude a neurological cause. He said that if Zeke did not have a family history of epilepsy he would place less weight on the possibility of a neurological cause but he could not discount it.

Sudden Cardiac Death:

62. Sudden cardiac death is a more common cause of death in children than SUDEP. It most commonly occurs to people in sleep or rest. If I were to ascribe a cause of death on statistical analysis alone, this cause would be considerably more likely. However, the cardiologist experts agreed that there was insufficient evidence to conclude that Zeke’s death was from a cardiac cause.

[3] Sudden cardiac death is also extremely rare in the age group 6-10, Semsarian et al “A Prospective Study of Sudden Cardiac Death among Children and Young Adults” 2016 New England Journal of Medicine p2445.

63. It was common ground between all of the experts that the Patent Foramen Ovale in Zeke's heart was not a cause or contributing cause of death (because there was no evidence of a large embolic cerebral infarction at autopsy or any evidence of an embolism in tests conducted prior to death).

64. The cardiologists did not place the same weight on the ECG result suggesting a ventricular conduction delay as the pathologist Professor Thomas. Professor Thomas opined that an incomplete right bundle branch block could have been a cause of death because he had seen adults with that condition who had died suddenly from no other apparent cause.[4] He had not seen or read about a child's death being ascribed to incomplete right bundle branch block. Dr Roberts stated that an ECG reading suggesting a ventricular conduction delay (or incomplete right bundle branch block) was a normal variant in children as did Professor Cheung in his report. Professor Semsarian agreed, noting in the conclave that people can die suffering from common conditions without those conditions being responsible for the death. The cardiologists did not believe the ECG result provided any assistance in assigning a cause of death.

65. The cardiologists also agreed that the variation between the measurement of the left ventricular free wall between the echocardiogram (7mm) and autopsy (14mm) was not of significance in determining the cause of death. They agreed with Professor Du Flou that variation was expected between measurements taken in life and after death. Professor Thomas' evidence was also to the effect that he considered the variation to be unexceptional. Professor Semsarian agreed with Professor Thomas that one would not expect significant hypertrophy to develop in four months.

66. The cardiologists also agreed that the post-mortem measurement was not of itself proof of cardio-myopathy, although that was a possible explanation. While the measurement was at the upper limit for Zeke's age range, Dr Roberts stated it was not abnormal for a boy of his size and weight (50.5 kg and 143cm). He said heart size is proportional to body size. Dr Roberts deferred to the expertise of Professor Du Flou who stated in his report that "the heart size was normal and the ventricular wall dimensions were probably on the high end of normal".

67. The inflammation detected at autopsy in Zeke's heart is of more significance. It was agreed by the experts that the finding was abnormal, not incidental. The finding supports the possibility that Zeke died from sudden cardiac death. But the finding is not determinative. Dr Roberts stated that the inflammation could have been caused by a viral condition unrelated to the cause of death (he also said such a virus would not necessarily show any obvious symptoms before death). Professor Thomas also accepted the possibility that viral infection caused the inflammation detected in Zeke's heart (and in his brain and other parts of his

[4] There was no pathological evidence of any problem in the conduction system of the heart because investigation of that possibility would require serial sectioning of the conduction system. This practice does not occur in NSW.

body). Professor Semsarian stated that the heart abnormalities pointed towards a cardiac event as a cause of death but were not determinative. He agreed that some of the pathological changes in the heart had also been seen in cases of SUDEP. Dr Lawson considered the post-mortem changes outside his expertise but noted the findings, to his knowledge, were equally suggestive of SUDEP and SCD.

The Deletion of Echocardiogram Images:

68. After Zeke died Mrs Howard made a request for the echocardiogram conducted by Dr David Murphy on 25 May 2011.

69. It was then discovered in December 2011 that the echocardiogram images had been permanently deleted along with two month's worth of other echocardiogram results. The images of at least 151 patients were lost and were irretrievable.

70. A forensic investigation was conducted by consulting service Lyonswood in October 2012. It concluded that the images had been deleted in two batches. Both dates were before Zeke died – some images were deleted on 12 August 2011 and the remaining images on 26 August 2011.

71. Lyonswood found that the log-in used belonged to Dr David Murphy, however that log-in was a default log-in for all doctors. A comparison of attendance records with the dates of deletion suggests that the images were actually deleted by Dr Stephen Cooper. Dr Cooper voluntarily spoke to police investigating the death and confirmed that he may have deleted the images. Dr Cooper was a visiting paediatric cardiologist who used a room and equipment at Nepean Neo-Natal Intensive Care as part of an informal arrangement with staff at Nepean Hospital. He understood that stored images on the ultrasound machine were downloaded and archived by a man called Murray approximately every month when the hard drive on the machine became full. Dr Cooper said the images would build up on the hard drive and it would get so full he would not be able to save images after he conducted a test. As a result, when the hard drive of the machine was full he had developed a practice of deleting the oldest images to free up some space. He assumed that the images he deleted had already been archived. I consider that it was reasonable for Dr Cooper to assume the Hospital had a functioning archiving system for equipment used in the neo-natal unit. But I note that Dr Cooper said he deeply regretted that he did not check the images were archived before he deleted the images and that he had personally implemented a new system for image storage to make sure it never happened again.

72. Murray Hinder confirmed he was employed at Nepean Hospital from 2001 to July 2011 as a biomedical technical officer. As part of his role in maintaining medical equipment he would download the data on the ultrasound machine onto a hospital network. He said the system, if one could call it that, was that he would download images if the machine was placed in his workplace with a note on a

paper towel saying the hard drive was full. Murray Hinder was not aware of any formal policy or procedure for routine back up or for the deletion of data.

73. The evidence at the inquest confirmed that there was no system in place. In particular there was no requirement for Mr Hinder to confirm a back-up had taken place in written form or to free up the hard drive of the archived images once the back-up process had occurred.

74. The deletion of Zeke's echocardiogram, along with medical images belonging to 150 other patients, was preventable. It has compounded the tragedy in this case. It has caused immense distress to Zeke's family.

75. Original medical images of investigative tests are extremely important information. The inquest heard that treating physicians use the images to inform medical treatment. They sometimes return to archived images to treat chronic problems or to reinvestigate a diagnosis. Coroners may need to refer to archived images to investigate the cause of death. The deletion of the echocardiogram could have deprived this inquest of critical information about the manner and cause of Zeke's death. In the present case Professor Semsarian said that the deleted echocardiogram was the one original image he would have had reference to in giving an opinion as to the cause of death. One can only imagine the distress this evidence must have caused Zeke's family.

76. Every Local Health District has an obligation to ensure that they have robust and rigorous systems to keep the medical data stored on their equipment safe and secure. This did not occur at Nepean Hospital in 2011. The system for retention and storage of data captured by the Vivid I ultrasound machine was seriously inadequate. Mr Hinrichsen conceded this to be the case when he gave evidence on behalf of the Nepean Blue Mountains Local Health District (NHD). His concession was responsible and appropriate. It has assisted the inquest and saved valuable court time and resources. This is to be commended.

77. However, I agree with Counsel Assisting that it must be observed that the immediate response by the Nepean Hospital to the deletion of the images was also inadequate. The discovery of the loss of thousands of medical images belonging to 151 patients should have been of immense concern to the Local Health District. The loss of those images demonstrated that the data stored on the machines was vulnerable to inadvertent deletion. Most of the data belonged to patients of the visiting clinicians but it may also have included images belonging to neo-natal intensive patients of Nepean Hospital. The Nepean Hospital should have taken immediate steps to eliminate the risk that data belonging to extremely vulnerable babies under their care could be destroyed.

The steps taken by the NHD should have included:

78. First, a prompt and thorough investigation of how and why the data was deleted. This investigation should have commenced and concluded well before October 2012 when the NHD engaged Lyonswood Consulting. It does not appear that the identity of the person who deleted the information was known until November 2012. The tardiness in the investigative response unfairly created a cloud of suspicion over Dr Murphy after Zeke died unexpectedly. It is not clear what investigation would have occurred if the Howard family had not publically and vigorously pursued the issue.

79. Secondly, the NHD should have immediately clarified in writing with the visiting clinicians at the hospital:

a) who had primary responsibility for the storage and retention of the data;

b) in what circumstances, if any, data should be deleted; and

c) if deletion was permitted, how clinicians could ensure that the data was archived on the hospital system through reference to an accessible written document.

80. Until those steps were taken there was a risk that a clinician could irreversibly delete original images if the ultrasound hard drive became too full to store medical images. That risk was unacceptable. There was no evidence to suggest the steps outlined above could not have been taken by the NHD.

81. The clarification of data retention arrangements set out in Mr Hinrichsen's letter to visiting clinicians on 29 November 2012 was a step, albeit a late one, in the right direction. The letter still failed to implement a clear system for data storage and retention. There were no instructions about when data could or should be deleted. There was no information about how clinicians could ascertain if data stored on the hard drive had been archived in the hospital system.

82. From September 2011 to September 2014 the safety of the data was dependent upon the visiting clinicians saving the images on a disk or flash drive (a step they were not requested to take until 29 November 2012) and upon the relevant staff at Nepean Hospital (two busy staff specialists and a senior bio-technical officer) remembering to periodically back up the data. No one person at Nepean Hospital was given this responsibility. There was no instruction as to precisely when back-up should occur. There was no instruction to record the date spectrum of the back-ups in an accessible written form.

83. There is no evidence to suggest that any images were deleted after August 2011 and the inquest was informed that some investigation has taken place to confirm that no further images have been lost.

84. The inquest was informed that "in about September 2014" Nepean Neonatal Intensive Care Unit purchased a new ultrasound machine. This machine automatically archives images onto the hospital network. The machine also has a back-up system whereby the machine transmits images wirelessly if the machine

is not connected to a data port. Furthermore, an automatic report is generated if there is any problem with the transmission of data so that staff are immediately made aware if there has been a problem archiving images. The inquest also heard evidence about the system of storage and retention of other images.

85. In the circumstances, it is not necessary to make any recommendation arising from this case. The circumstances of the deletion and the response have been addressed in my findings in order to emphasise the importance of retention of medical images which can be of such importance to the Coronial jurisdiction.

Manner and Cause of Death:

86. The finding of the manner and cause of death is ultimately a legal question. The evidence of experts informs the decision. In this inquest I have had the benefit of the combined expertise of pathologists, clinicians (both paediatric and adult) and medical academic expertise through the research work of Professor Semsarian and Professor Du Flou.

87. I have reached the conclusion that the evidence does not allow me to make a determination as to the manner and cause of Zeke's tragic sudden death. This is an unusual case because I make that finding despite fully accepting that of the two major possibilities, that is death related to undiagnosed epilepsy and death from a cardiac condition, a death from a cardiac condition is more likely. While the role of a Coroner is to determine the cause of death "on the balance of probabilities" I am of the opinion that an expression of likelihood in this case could be misleading.

88. My reasons are as follows:

The weight of the expert evidence:

89. All of the experts except Professor Thomas were of the opinion that the cause of death was best described as "unascertained" or "undetermined". Professor Semsarian expressed that opinion particularly forcefully in the conclave. Professor Semsarian has had the advantage of the combined expertise of pathologists, clinicians and geneticists in his two large research studies on Sudden Cardiac Death and Sudden Death in Epilepsy. He said as many as 40% of sudden deaths of young persons fall into this category.

90. Mr Segal urged me to accept the evidence of Professor Thomas, partly on the basis that Professor Thomas had the most expertise in determining cause of death and partly by emphasising the evidentiary features pointing towards possible cardiac conditions. I do not consider I have enough information to conclude that Professor Thomas has greater expertise than the pathologists Dr Bailey and Dr Du Flou, who also gave evidence but were not required for cross-examination. Further, Professor Thomas accepted that the death was best

described as “unascertained” on a purely pathological basis. I do not consider that the difference between the way clinicians and pathologists might ascribe a cause of death means I should give the evidence of the clinicians less weight in this case.

91. Counsel Assisting notes that Professor Thomas’ expertise had limitations. He had no experience in examining children who died of epilepsy. His understanding of SUDEP was based solely on medical literature and accordingly was inferior to both Dr Lawson and Professor Semsarian. He accepted that paediatric clinicians would have greater expertise on the significance of the neurological symptoms experienced by Zeke during life. Finally, he placed some weight on the outcome of genetic testing but accepted he had no expertise in the area. Professor Semsarian did have such expertise and stated in the conclave that the genetic tests provided no assistance in determining Zeke’s death.

92. Mr Segal also submitted that Dr Phillip Kolos, the paediatrician who saw Zeke on 6 and 7 May 2011, favoured a cardiac cause in differential diagnosis and his opinion should be given weight as he examined Zeke. Dr Kolos did not give any opinion about the cause of death in the material provided to the inquest, he did not give evidence at the inquest and Mr Segal did not request that he be examined. With the greatest respect, I cannot place much weight on Dr Kolos’ initial diagnostic inclination because I do not know if he maintains his view in light of the post-mortem findings and other evidence presented to the inquest.

93. The experts in the conclave were resistant to evaluating whether individual findings were more suggestive of one possible underlying cause or another. Each of the experts gave evidence to the effect that the complete clinical picture was more important than individual pieces of evidence and the overall conclusion should be that the death was unascertained. I accept their evidence.

The absence of complete information.

94. There is insufficient information from which I can conclude a cause of death in circumstances where the indicia of two major possible causes are essentially indistinguishable and the evidence is incomplete. I accept that statistically it would be highly unusual for Zeke to die from undiagnosed epilepsy. But I am extremely reluctant to make a finding that would be considered to exclude such a possibility on the basis of statistics. As I noted above, Zeke’s sudden death was exceptionally unusual for any cause. Zeke had symptoms which a paediatric neurologist specialising in childhood epilepsy considered likely to be undiagnosed focal epilepsy. Epilepsy in children is best diagnosed by clinical examination by a specialist paediatric neurologist, ideally supplemented by witness accounts or videos of seizures and an EEG. This did not occur before Zeke died. I am not critical of any party for the absence of evidence but, to be trite, absence of evidence is not evidence of absence. I have more evidence about the possibility of a cardiac event partly because more cardiac investigations were conducted.

Only one epilepsy gene of many was investigated and there is no reliable evidence about family history.

95. Even if Zeke did have a diagnosis of epilepsy there would remain the question of whether he had a misdiagnosed heart condition mimicking epilepsy. I agree that it is speculative to suggest that an epilepsy diagnosis would have prevented Zeke's death. In fact, there is some evidence that treatment for epilepsy can be a risk factor for SUDEP[5]. My understanding of SUDEP and SCD is that there is much to be discovered and the possible inter-relationship between the phenomena is still being explored. In these circumstances, I consider a finding of sudden unexpected death to be the only responsible finding that I can make on the available evidence.

96. I would like to thank the officer in charge for their investigation.

97. I would also like to thank Counsel Assisting, Ms Kirsten Edwards, whose submissions I have largely adopted in these findings. I would also like to thank her instructing solicitor, Ms Janet de Castro Lopo from the Office of General Counsel.

98. Finally, I would like to pass on my heartfelt condolences to Sonja and William Howard and their family. They have persisted in their quest to try and find out what happened to their much loved little boy, Zeke. I would like to thank them for their participation in this process. It has been long and hard and at times frustrating for them. Their efforts have done much to bring together all that is known about the tragic phenomenon of sudden and unexplained death in the young. The body of expert knowledge that has been compiled for this inquest may well help other families and will certainly forge the way for more to be done by way of prevention.

Findings required by s81(1):

As a result of considering all of the documentary evidence and the oral evidence heard at the inquest, I am able to confirm that the death occurred and make the following findings in relation to it:

The identity of the deceased

The deceased person was Ezekiel Howard.

Date of death

He died on 12 September 2011.

Place of death

He died at Colo Heights, NSW.

[5] Tomson et al, Sudden Unexpected Death in Epilepsy: A Review of Incidence and Risk Factors, Epilepsia 46 (Suppl.11) 45-61, 2005, 55-56; Nouri, et al, Sudden Unexpected Death in Epilepsy: Overview, Autopsy Findings, E-medicine, Medscape, December 2015 at p7-9/15.

Manner and Cause of death

The death was sudden and unexpected. The evidence does not allow me to identify the cause of death.

I close this inquest.

Magistrate Teresa O'Sullivan

Deputy State Coroner

Date: 8 December 2017