Coroners Act 1996
[Section 26(1)]

Coroner's Court of Western Australia

RECORD OF INVESTIGATION INTO DEATH

Ref: 39 /19

I, Sarah Helen Linton, Coroner, having investigated the death of ***Dylan James RILEY*** *with an inquest held at the* ***Perth Coroner's Court, Court 51, CLC Building, 501 Hay Street, Perth*** *on* ***29 August 2019*** *find that the identity of the deceased person was* ***Dylan James RILEY*** *and that death occurred on* ***1 August 2015*** *at* ***Fiona Stanley Hospital, Murdoch****, as a result of* ***pulmonary embolism secondary to nephrotic syndrome*** *in the following circumstances:*

Counsel Appearing:

Mr D Jones assisting the Coroner.
Mr J Carroll (State Solicitor's Office) appearing on behalf of the South Metropolitan Health Service, Dr Willis and Dr Swaminathan.

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INTRODUCTION

1. Dylan Riley was born in 1999. He was diagnosed with paediatric nephrotic syndrome in 2005, when he was six years old, and received medical treatment for his condition. He responded well to treatment and his long-term outlook appeared good. It was considered very possible that Dylan might outgrow the condition as he moved towards adulthood, as the majority of children with his type of nephrotic syndrome do.

2. Dylan had frequent relapses over the years, but responded well to steroid treatment each time. After a change in medication in January 2015, he remained in remission until early July 2015. When his symptoms began to re-emerge, it did not generate concern, as the medication he was receiving usually wears off after about 6 to 12 months and he was due for another dose. Dylan was started on steroid treatment and a plan was put in place to give him another dose of his medication when he had come back into remission.

3. Dylan became increasingly unwell on 29 and 30 July 2015, with significant fluid retention and increasing shortness of breath, and he was taken to hospital by his parents on 31 July 2015 for further medical review. Dylan was noted to be short of breath and was hypoxic, tachycardic and hypotensive. He was given fluids and a prophylactic dose of an anticoagulant medication, Clexane, as he was known to be at increased risk of thrombosis due to his condition. He was then transferred to Fiona Stanley Hospital (FSH) for further treatment that night.

4. After further investigations, it was felt that Dylan's symptoms were related to acute pulmonary oedema (fluid on the lungs) although a pulmonary embolism remained part of the differential diagnosis. Although he was given a further dose of Clexane overnight, no specific investigations for a pulmonary embolism were performed. Treatment for acute pulmonary oedema was given and he was transferred to ICU.

5. At about 7.00 am on 1 August 2015 Dylan's oxygen saturations dropped and he became increasingly unwell and confused. Just before 10.00 am a bedside echocardiogram showed right heart failure and a diagnosis of probable massive pulmonary embolus was made. A cardiothoracic surgeon was consulted, who advised that Dylan was not a candidate for surgical removal of emboli and suggested thrombolysis (breakdown of blood clots with medication) instead. He was given a thrombolytic agent but sadly his condition did not improve. Dylan went into cardiac arrest and couldn't be resuscitated. Dylan died at 12.25 pm on 1 August 2015. He was only 16 years old.

6. A doctor completed a medical certificate indicating the cause of death was pulmonary embolism on the background of his known nephrotic syndrome.[1]

7. At the time of Dylan's passing, his mother, Jennifer Murphy, and her partner, Wesley Riley, decided not to pursue an autopsy and coronial

[1] Exhibit 1, Tab 2.

investigation. Ms Murphy says they made that decision on advice from staff at FSH. Dylan's death was deemed by medical staff not to be a coroner's case. A death certificate was issued by a doctor from FSH and the death was not reported to the coroner.[2]

8. After Ms Murphy and Mr Riley had recovered from the initial shock of Dylan's sudden death, they began to have concerns about Dylan's treatment in hospital and also became concerned that his death may have been preventable. Ms Murphy and Mr Riley made contact with staff from the Office of the State Coroner, and then wrote to the State Coroner on 20 September 2015 requesting a coronial investigation be undertaken into Dylan's death.[3]

9. An investigation was commenced in October 2015, which prompted requests for Dylan's medical records and then a number of medical expert reports. Some of these reports took time to obtain, and they prompted other lines of enquiry. When all the medical information had been obtained, the matter was reviewed and ultimately it was decided that an inquest should be held to further explore why the pulmonary embolism was not diagnosed at an earlier stage, and whether Dylan's death was preventable.

10. I held an inquest in Perth on 29 August 2019. Evidence was heard from Dylan's treating Paediatric Nephrologist, Dr Francis Willis and the Head of Nephrology at FSH, Dr Ramyasuda Swaminathan. In addition, Dr Ram Tampi, a Clinical Haematologist, reviewed Dylan's case and provided an expert opinion on his medical treatment and cause of death. I also had before me detailed statements from doctors involved in Dylan's care prior to his death and his medical records and information provided by his family, forming two volumes of exhibits.[4]

BACKGROUND

11. Nephrotic syndrome is a condition which affects the kidneys and results from damage to the glomeruli, which are the tiny blood vessels that filter waste and excess water from the blood and make urine. When a person has nephrotic syndrome, the kidneys leak a large amount of protein into the urine (proteinuria) and this causes a low level of protein in the blood. Low levels of protein in the blood, lowers the osmotic pressure in the blood vessels and allows fluid to leach out of the blood vessels into the surrounding tissues. This results in swelling and fluid retention, known as oedema. Nephrotic syndrome can also cause high blood levels of cholesterol and other fats (lipids) and an increased risk of developing infections, due primarily to losing antibodies in the urine. Relevantly to Dylan's death, patients with nephrotic syndrome are also at an increased risk for thrombotic events such as deep venous thrombosis, renal vein thrombosis and pulmonary embolism, although I am told these complications are rare in children.

[2] Exhibit 1, Tab 3.
[3] Exhibit 1, Tab 3.
[4] Exhibits 1 and 2.

12. Dylan was diagnosed with childhood nephrotic syndrome in 2005. After being diagnosed, Dylan's nephrotic syndrome was managed by Paediatric Nephrologist Dr Francis Willis until Dylan's unexpected death in August 2015.[5]

13. Nephrotic syndrome is a description of a series of signs and symptoms for which there are multiple causes.[6] Dylan's nephrotic syndrome was believed to be due to minimal change disease, which is one of the most common causes of nephrotic syndrome in childhood and carries a generally good prognosis.[7] The two factors that best predict long-term outcome in minimal change nephrotic syndrome are the age of onset and response to steroids. Dylan presented in the typical age group and responded very well to steroids on all occasions, so this put him in the good long-term outlook group.[8]

14. Dr Willis gave evidence that the vast majority of paediatric patients grow out of it as they reach adulthood, with one study finding only one in 20 have continuing disease in their mid-20's. The longer the patient goes without growing out of it, the less likely they will do so, but Dr Willis indicated that there was still some possibility that Dylan would outgrow the disease. Dr Willis explained that doctors are still uncertain as to why paediatric patients outgrow the disease, with one possibility being the changes the body undergoes in puberty, so when or if Dylan might grow out of it couldn't be predicted.[9]

15. In the meantime, steps were being taken prior to Dylan's death to transition him into adult care as he was finishing school, looking at starting an apprenticeship and moving into adulthood. He still required ongoing treatment at that stage and it couldn't be predicted if that would change.[10] Long-term, if Dylan continued to suffer from nephrotic illness, it could lead to complications such as renal failure, but for his entire illness Dylan had normal kidney function, so there was no suggestion he might reach that stage anytime soon.[11]

16. Dylan was initially treated with standard steroid treatment, to which he responded. However, he had frequent relapses, which led him to be classified as a frequent relapser (which is defined as four or more relapses in a 12 month period).[12] Because of his frequent relapses he had a kidney biopsy, the results of which were consistent with Minimal Change Disease, as expected.

17. The frequency of Dylan's relapses was not a concern to his doctors in terms of his prognosis, as Dylan was steroid responsive on each occasion. However, it did require a change to a steroid sparing agent.[13]

[5] Exhibit 1, Tab 6.
[6] T 7; Exhibit 1, Tab 6B.
[7] Exhibit 1, Tab 4 and Tab 6.
[8] T 7.
[9] T 8 – 9.
[10] T 8.
[11] T 8.
[12] T 10 – 11; Exhibit 1, Tab 6.
[13] T 10 - 11.

18. Dylan was trialled on a medication called levamisole, which has advantages in terms of side effects, as it is very benign but has the disadvantage that it doesn't work in the majority of cases. It didn't work in Dylan's case.[14]

19. Dylan was then started on an alternative, and more reliable, steroid sparing agent called Tacrolimus. He responded well to Tacrolimus and went several years with either no further nephrotic relapses, or only one or two nephrotic relapses. All of the relapses responded to standard steroid treatment. This remained the case for approximately eight years until 2014, while Dylan continued on Tacrolimus.[15]

20. However, in 2014 Dylan had three relapses in May, August and November. The relapse in November occurred while the steroid treatment was being tapered. On each occasion he responded to standard steroid treatment, but it took up to three weeks for him to do so. It was noted that Dylan had been on Tacrolimus for some time and it was felt that Dylan may have outgrown his current dose. There was talk of increasing his dose. When he was reviewed in December 2014 Dylan was well and in remission, so Tacrolimus was continued.[16]

21. Dylan relapsed again in January 2015, which reinforced his categorisation as a frequent relapse. He went into remission after about one week of standard steroids. At this stage, Dylan's treatment was changed to another medication called rituximab, which has the advantage that if it works it will give the patient several months without needing to take medications. The effect eventually wears off over a 6 to 12 month period, and the patient will generally require re-dosing, but it still provides a lengthy period without requiring other medication.[17] Dylan was administered intravenous rituximab with good effect. He was then able to come off the Tacrolimus.[18]

22. Dylan remained in remission for approximately six months from this time. Plans were made for his transition to adult medical care at Rockingham General Hospital, which was closer to his home, and FSH as his tertiary care hospital, replacing the Perth Children's Hospital (PCH). Dylan was in Year 11 at this stage and it was planned for the transition in care to take place when he finished the school year. It was expected his treatment would have been reviewed on his transition to a new adult treating team.[19] Dr Willis explained in his evidence that he works from both PCH and FSH, so he was able to start seeing Dylan at the FSH clinic, to help him to become familiar with the hospital and transition to adult care gradually.[20] Dr Willis described Dylan as an intelligent young man with really good family supports. Dylan and his family were relatively well-informed about his condition and he was very sensible and cooperative with his treatment regime, all of which indicated his transition to adult care should go well.[21]

[14] T 11.
[15] Exhibit 1, Tab 6.
[16] Exhibit 1, Tab 6.
[17] T 11.
[18] Exhibit 1, Tab 6.
[19] Exhibit 1, Tab 6.
[20] T 12.
[21] T 12 – 13, 22.

23. Dylan remained in remission until early July 2015, when he developed protein in his urine, lethargy, swollen feet and abdominal pain. The timing was consistent with the rituximab effect beginning to wear off, and also coincided with symptoms of gastroenteritis. Dylan was restarted on the steroid treatment prednisolone.[22]

24. He commenced the course of prednisolone and was reviewed by Dr Willis at the FSH clinic four days later, on 27 July 2015. Dylan looked well at that time, but had signs of relapse. Dylan was started on the diuretic frusemide for his oedema and the antibiotic Keflex.[23] Dylan was also rebooked for a further clinic appointment the following week with a plan made that if he had not gone into remission he would then be admitted to hospital so he could be given intravenous steroids.[24] Once Dylan was back into remission, it was planned to re-treat him with rituximab, in the hope it would give him another six months or more of control of his symptoms.[25]

25. Dr Willis did not believe Dylan showed any sign of thrombosis at the time he saw him at the clinic in late July 2015, although Dr Willis accepted sometimes the signs can be very subtle, to the point of being undetectable, so it could not be ruled out entirely.[26]

26. Dylan's relapse worsened after the clinic visit despite already being on steroid treatment. Dr Willis indicated the steroids typically take a period of time to work, ranging on average from 7 to 14 days, so it was not necessarily an indicator that he had become steroid resistant. However, Dylan's quick deterioration did surprise him.[27]

27. Over the course of the following week, Dylan remained home from school, although he was able to assist with household chores and walk his younger sister home from school. On Wednesday, 29 July 2015, Dylan told his mother he felt dizzy and tired after collecting his sister, although he recovered quickly and was still eating and drinking normally. The following day he stayed on the couch, watching television, for most of the day and complained of feeling short of breath when walking. Dylan's symptoms worsened further the next day. When his parents observed him having increased work of breathing and a colour change, they immediately took him to the Emergency Department of Rockingham Hospital.[28]

PRESENTATION TO ROCKINGHAM HOSPITAL

28. Dylan arrived at Rockingham Hospital just after 6.00 pm on 31 July 2015. His parents noted the Emergency Department was excessively busy, and they had to wait about half an hour before being triaged, and then another half an hour to be seen by a doctor.[29]

[22] Exhibit 1, Tab 6.
[23] Exhibit 1, Tab 3.
[24] T 16.
[25] T 16.
[26] T 18.
[27] T 17.
[28] Exhibit 1, Tab 3.
[29] Exhibit 1, Tab 3.

29. Dylan was seen by Dr Hughes. Dylan's medical history was noted and that he had recently commenced prednisolone, a diuretic and antibiotics due to a relapse of his nephrotic syndrome. It was recorded that he felt short of breath and had put on 10kg in weight in the last 10 days, which was likely to be fluid related.[30] Dylan was noted to be pale, his blood pressure was low at 85/70 and his heart rate was increased at 105 bpm. His oxygen saturations were low, at 88% when walking and 93% at rest, but they later increased to 100%. A few crackles in his lungs were noted, as well as bilateral pedal oedema (swelling of his feet). A urine test showed a large amount of protein, blood, leucocytes and nitrite. A blood gas analysis and various blood tests were also performed. Following these investigations, Dylan was diagnosed with a relapse of nephrotic syndrome.

30. Dr Willis was not on duty that evening, but he received a call on his mobile telephone from a doctor in the ED at Rockingham Hospital informing him that Dylan was unwell. Dr Willis believes he gave an outline of Dylan's case and suggested he be transferred to the adult renal team at FSH. Dr Willis felt this to be a more appropriate setting than a children's hospital for Dylan, where equipment and staff are set up for smaller individuals than him. As noted earlier, a plan was also already in place to transition his ongoing tertiary care to that hospital.[31]

31. After that discussion, and after discussion with a Princess Margaret Hospital Paediatric Nephrology Consultant who gave similar advice, Dylan was commenced on a normal saline infusion and arrangements were made for him to be transferred to FSH. In view of his age, there were some issues determining whether he should be admitted under the paediatric or adult renal team, but he was eventually accepted under the adult team.[32]

32. Other than that initial call from Rockingham Hospital, Dr Willis indicated that he did not receive any further calls about Dylan's care. He also indicated he would not have expected the renal or intensive care teams at FSH to contact him for advice. Dr Willis explained that these are his colleagues and they are specialists and experts in their own right and are appropriately trained to manage young people such as Dylan without his assistance.[33]

33. At 7.53 pm Dylan complained of central chest pain of 5/10 severity. An ECG was performed, which showed sinus tachycardia. A chest x-ray showed a small pleural effusion (fluid around the lung) at the right lung base.[34] According to the medication chart, Dylan was given two panadeine for pain relief at 8.15 pm.

34. At 8.21 pm the observation chart recorded an increased pain score of 7/10, despite the analgesia given. He was given enoxaparin (Clexane) 40 mg subcutaneously, which was a prophylactic dose of anticoagulant for

[30] T 19.
[31] Exhibit 1, Tab 6.
[32] Exhibit 1, Tab 6.
[33] Exhibit 1, Tab 6.
[34] T 58.

prevention of thromboembolism. He left Rockingham Hospital for transfer by ambulance to FSH Emergency Department (ED) at 9.22 pm.

FIONA STANLEY HOSPITAL ADMISSION

35. Dylan arrived at FSH ED at 9.57 pm. He was allocated a Triage Code of 2. He was noted to be pale with rapid, laboured breathing and a rapid pulse.[35] Although these symptoms could raise suspicion of a pulmonary embolism, Dr Willis gave evidence that they were statistically more likely to be indicative of a pulmonary oedema, which involves fluid within the lungs that causes respiratory symptoms, and is a common feature associated with nephrotic syndrome.[36]

36. Dylan was seen by the Renal Registrar, Dr Nyi Aung, at 10.44 pm. Dr Aung had not met Dylan prior to that evening. Dr Aung reviewed Dylan in the presence of his mother and a fully typed Medical Admission form was then completed. Dr Aung reviewed Dylan's test results and conducted a physical examination.[37]

37. It was noted that Dylan's heart rate was between 120 – 140 bpm and his oxygen saturations were low at 91%. He also had moderate oedema. An ECG was reported to show sinus rhythm with no acute changes and a chest x-ray was reported to show "feature of APO (acute pulmonary oedema)" and no cardiomegaly (enlarged heart). The provisional diagnosis was fluid overload with intravascular volume depletion on the background of relapsing idiopathic nephrotic syndrome.[38] Importantly, note was made of Dylan's increased risk of thrombosis, for which he was prescribed prophylactic anticoagulation. Dr Aung did not think that Dylan had a pulmonary embolism at the time that he was reviewed but was aware it was a possible risk factor.[39]

38. The management plan included:

    - Admission under Renal Physician Dr Brian Siva;
    - Daily weights and commencement of an input/output chart;
    - Continue prednisolone 40mg twice daily;
    - Increase frusemide to 80mg twice daily;
    - Prophylactic Clexane 40mg daily;
    - Cardiac monitor bed;
    - IV albumin infusion; and
    - Non-invasive ventilation.

39. Dr Siva was on call but not in the hospital, and Dylan's case was discussed with him by Dr Aung over the phone. Dr Siva did not recall the specific conversation, but noted that nothing in the plan causes him to think that he did not agree with it. It was intended that Dr Siva would review Dylan on

[35] Exhibit 1, Tab 3.
[36] T 19.
[37] Exhibit 1, Tab 10 [20].
[38] Exhibit 1, Tab 10 [26].
[39] Exhibit 1, Tab 10 [28] – [29].

post-intake rounds the next day to review the treatment plan further, but Dylan was already in ICU and critically unwell by that stage.[40]

40. It is clear Dr Aung considered the possibility of thromboembolic disease and was aware that Dylan was at risk of pulmonary embolus, although it was not his primary diagnosis. Dr Aung ordered a further dose of Clexane to be administered for that reason.

41. Dylan was administered the diuretic frusemide twice that night. An entry at 10.52 by ED Registrar Dr Helen Jones recorded that Dylan was "hypoxic taccycardic (sic) and hypotensive." She reported the chest x-ray as "not overtly in APO." Her impression was that Dylan was intravascularly depleted but documented her opinion that "Fluid balance is going to be very difficult."

42. An entry at 12.39 am on 1 August 2015 by Dr Aung recorded that Dylan had a good urine output after the frusemide doses but his heart rate was still increased, his oxygen saturations were only 91% on 2 litres of oxygen and he had increased work breathing. He was commenced on CPAP and arrangements were made for him to be admitted to the ICU. This was because Dylan needed a cardiac monitored bed and further management and the Acute Medical Unit could not support him, so the ICU was the most appropriate place. The Renal Ward does not have cardiac monitored beds so it was not an option.[41]

43. The ED observation charts show that at 1.10 am Dylan's oxygen saturation dropped to 59% while on CPAP and he became agitated. Evidence was given at the inquest that this could be consistent with pulmonary oedema.[42]

44. Dylan was admitted to the ICU at 1.50 am on 1 August 2015. At that time his blood pressure was 130/60, his oxygen saturation was 98% on high flow nasal prongs and his respiratory rate was still increased at 25 – 30 breaths per minute and he had an elevated heart rate throughout the night. However, subjectively, he indicated he felt more comfortable with his breathing.[43]

45. It was recorded on the completed medication summary report that Dylan was given a further dose of 60mg Clexane overnight, but it is not clear at what time this was administered.

46. According to the progress notes, between 7.05 and 7.20 am Dylan's oxygen saturations dropped and he became increasingly confused. He developed rapidly increasing left should pain that radiated across his chest. He was commenced on fentanyl, a strong opioid pain relief, at 8.45 am. His condition worsened and he became haemodynamically unstable. An entry in the medical progress notes by the ICU RMO, Dr Tsang, at 9.43 am reported a bedside echocardiogram showed right heart failure and a diagnosis of probable massive pulmonary embolus was made.

[40] Exhibit 1, Tab 9 [24] – [28].
[41] Exhibit 1, Tab 10 [34] – [35].
[42] T 39.
[43] Exhibit 2, Tab 4.3 [158]

47. Cardiothoracic surgeon Mr Gilfillan was contacted and advised that Dylan was not suitable for embolectomy (the surgical removal of emboli) but suggested thrombolysis (breakdown of clots with medication). A diagnostic CT Pulmonary Angiogram (CTPA), which is a test that can identify a clot, was not done due to Dylan's severe haemodynamic compromise and there was sufficient evidence on the echocardiogram to support the diagnosis of massive pulmonary embolism.

48. The medication report stated that Dylan was given the thrombolytic agent Aletaplase 50mg. Despite the attempt at thrombolysis, Dylan's condition did not improve and he required increasing doses of noradrenaline, adrenaline and levosymendin.

49. The Cardiothoracic Surgical Registrar, Dr Skiba, recorded that Dylan was haemodynamically unstable and the decision was made to place him on ECMO (Extracorporeal membrane oxygenation) a treatment that uses a pump to circulate blood through an artificial lung back into the bloodstream. The medical staff experienced great difficulties accessing his veins, due to clots, and were unable to achieve ECMO, in spite of numerous attempts.

50. A transesophageal echocardiogram showed "significant clot burden" in the inferior vena cava. Dylan's condition further deteriorated and he went into cardiac arrest. Further attempts at resuscitation were unsuccessful and his death was confirmed by a doctor at 12.26 pm on 1 August 2015.

51. A death certificate was signed by Dr Zhou on 1 August 2015 identifying the cause of death as pulmonary embolism, with an approximate interval between its onset and death noted as two weeks. The antecedent cause was identified as nephrotic syndrome, which Dylan had been diagnosed with for many years.[44]

CAUSE AND MANNER OF DEATH

52. As noted above, the cause of death was given by a doctor as pulmonary embolism on a background of nephrotic syndrome. No post mortem examination was conducted, for the reasons set out below, so this is the only cause of death available. There was no suggestion in the evidence that there was any error in the cause of death given. Accordingly, I find that Dylan died as a result of a pulmonary embolism in the context of a relapse of his nephrotic syndrome. It follows that death occurred by way of natural causes.

REPORT TO THE CORONER

53. ICU Consultant Dr Ponasanapalli recorded a retrospective entry in the medical notes at 7.45 am on 3 August 2015, detailing that extensive discussions were held with Dylan's family and the coroner's office, and at that time it had been deemed that his death was not a reportable death.

[44] Exhibit 1, Tab 2.

54. On 19 August 2015 a family meeting was held with Dylan's parents, members of the FSH medical team and a senior social worker. It was explained to Dylan's family that the treating team initially got misled into believing that Dylan was 'intravascularly dry and extravascularly wet'. It was acknowledged that it was known that he was at high risk of developing a pulmonary embolism and had been given a prophylactic dose of Clexane initially and an additional dose of Clexane while he was in ICU, but no further investigations were undertaken to identify the cause of his persistent tachycardia and oxygen requirements. The medical team informed Dylan's parents that they were unsure whether the outcome would have been different if Dylan had received thrombolysis at an earlier stage.

55. As I noted at the start of the finding, Dylan's parents reflected after this meeting and made contact with counselling staff at the Office of the State Coroner to explore their options. On 20 September 2015 Dylan's parents wrote to the State Coroner and identified a number of questions they had about his medical care immediately prior to his death. They understood that the opportunity for a post mortem examination to be performed had passed, but asked for a coronial investigation to still be undertaken in relation to Dylan's death. They emphasised that their aim was not to try to lay blame but rather to ensure that, if errors or omissions were made, a coronial investigation might identify them and prevent a similar death occurring.[45]

56. In particular, Dylan's parents focus was on why they were not made aware of the possibility Dylan could develop thrombotic complications, why the doctors did not consider pulmonary embolism as a diagnosis sooner, and whether Dylan's death was preventable? Further, they wanted to ensure that any lessons that could be learned from Dylan's case be explored, to hopefully prevent the loss of another life in similar circumstances.[46]

PULMONARY EMBOLISM

57. Pulmonary emboli usually arise from thrombi clots that originate in the deep venous system of the lower extremities, such as the legs. However, they can also occasionally originate in the pelvic, renal, upper limb veins or the right heart chambers. The thrombi can travel through the circulation to the lungs. Large thrombi can lodge at the bifurcation of the main pulmonary artery or the lobar branches and cause haemodynamic compromise.

58. The classic presentation of pulmonary embolism is the abrupt onset of pleuritic chest pain, shortness of breath and low oxygen saturations. However, many patients with pulmonary embolism have no or minimal symptoms at presentation, and the symptoms can also be consistent with other conditions.

59. Pulmonary embolism is a medical emergency and a major cause of morbidity and mortality. Anticoagulation with heparin and warfarin remains the mainstay of treatment for pulmonary embolism. The primary objectives of

[45] Exhibit 1, Tab 3.
[46] Exhibit 1, Tab 3.

anticoagulant therapy are to diminish the propagation of the existing clot and to prevent recurrence. Thrombolysis to dissolve the clot, is also used to treat acute massive pulmonary embolism accompanied by haemodynamic instability or cardiogenic shock.

60. This is certainly not the first inquest in this State involving a late, or indeed missed, diagnosis of pulmonary embolism. A very relevant recent example is the inquest into the death of Kellie Anne Keogh.[47] In *Keogh*, the deceased was also diagnosed with nephrotic syndrome and died from a pulmonary thromboembolism. The inquest focused on the management of the deceased's risk of developing a deep vein thrombosis and pulmonary embolism and her anticoagulation therapy. Coroner Collins made a number of recommendations arising out of the evidence.

61. Relevant to this inquest, His Honour recommended that Sir Charles Gairdner Hospital consider developing guidelines or protocols for obtaining advice and guidance from consultant haematologists in relation to the management of patients at higher levels of risk of developing a deep vein thrombosis or pulmonary thromboembolism. Further, based on the expert opinion of Dr Tampi, who I refer to below, about a similar service in place at Royal Perth Hospital, Coroner Collins recommended that Sir Charles Gairdner Hospital consider developing a Thrombosis Management Service to provide specialist advice in relation to patients with an increased risk of deep vein thrombosis and pulmonary embolism.

62. In January 2015 I delivered a finding following an inquest into the death of another young woman from pulmonary embolism, Petra Zele. Ms Zele did not have nephrotic syndrome, but instead had an undiagnosed genetic mutation that increased her risk of pulmonary embolism, particularly when taken in conjunction with the contraceptive pill. The difficulty of diagnosing pulmonary embolism was discussed during the inquest and following her death Fremantle Hospital had implemented its own Diagnostic Pathway for pulmonary embolism, as well as targeting pulmonary embolism in education sessions for all medical staff.[48]

63. Dylan's case obviously has points of difference from the above cases, but there are also similarities in terms of the key questions of why a pulmonary embolism wasn't considered sooner and therapeutic anticoagulation commenced. What all of the cases demonstrate is the difficulty in diagnosing and then managing a pulmonary embolism, which in many cases can be fatal.

Risk of Dylan suffering a Pulmonary Embolism due to Nephrotic Syndrome

64. Dr Willis was aware of the cause of Dylan's death, namely a pulmonary embolism, when he provided a report and later gave evidence. Dr Willis' evidence was that such an occurrence in a paediatric patient with nephrotic syndrome was very, very rare and unexpected.

[47] Inquest into the death of Kelly Anne Keogh held 17 – 19 September and 16 October 2012, Coroner Collins.
[48] *Inquest into the death of Petra Zele, delivered 15 January 2015.*

65. Although there is general evidence that pulmonary embolism is a recognised complication associated with nephrotic syndrome, Dr Willis indicated that this is in the context of adult patients. Dr Willis explained that in paediatrics, pulmonary embolism in the context of nephrotic relapse is extremely rare. Dr Willis cited studies in support of this position and also his own personal experience of more than 30 years in medical practice. In those 30 years, Dr Willis gave evidence he had no personal experience of a pulmonary embolism in a young patient, and to the best of his knowledge nor have any of his nephrology colleagues at Princess Margaret Hospital.[49]

66. As to why the risk of venous thrombo-embolism may be greater in adult patients, Dr Willis suggested that it may be because adults have a different profile of nephrotic syndrome to children, including much greater relative numbers of Membranous and secondary nephrotic syndrome. In contrast, the dominant pathology for nephrotic syndrome in paediatrics is Minimal Change Disease, presenting as steroid sensitive nephrotic syndrome, where the overall risk for venous thrombo-embolism (VTE) or pulmonary embolism (PE) is much less.[50]

67. Dr Willis noted that Dylan's kidney biopsy in June 2006 suggested Minimal Change Disease and he had consistently responded to steroids, thus placing him in the relatively low risk group for a VTE or PE in the context of patients with nephrotic syndrome. Dr Willis did note that adolescent patients are at higher risk than younger children, and Dylan obviously fell into this category.[51] Still, the increase in risk was not such as to be considered significant.

68. Dr Willis noted that the increased prevalence of thrombosis and pulmonary embolisms in adult patients, meant that early diagnosis and management of a pulmonary embolism is probably more likely in an adult hospital than in a paediatric one, as it is a more common occurrence.[52] Therefore, the fact that Dylan had been sent to FSH rather than PCH probably improved his chances of an early diagnosis.

69. There are other risk factors for VTE or PE, but none of these factors were present with Dylan.[53]

70. Clinical features raising suspicion of VTE or PE in patients with nephrotic syndrome may include respiratory symptoms such as shortness of breath, raised respiratory rate, supplemental oxygen requirement or cardiovascular symptoms such as chest pain. These were all symptoms Dylan had. However, it is also important to note that pulmonary oedema, which is a much more common complication of nephrotic syndrome, can cause similar respiratory symptoms and signs.[54]

[49] Exhibit 1, Tab 6.
[50] Exhibit 1, Tab 6.
[51] T 17; Exhibit 1, Tab 6 and Tab 6B.
[52] Exhibit 1, Tab 6.
[53] Exhibit 1, Tab 6.
[54] Exhibit 1, Tab 6.

71. Dr Willis suggested it was not possible to know exactly when the onset of the pulmonary embolism occurred,[55] and this appeared to be agreed by the other experts.

72. Dr Willis believed there was nothing to suggest that Dylan had a PE at the time of any of his clinic reviews or at any other time, prior to his last hospital presentation. However, Dr Willis acknowledged that this is impossible to confirm, as PE may be mild and even asymptomatic, depending on its extent.[56] It follows therefore, that it is unlikely, but not impossible, that Dylan had a PE when he was reviewed in the clinic four days before he presented to hospital. However, it is much more likely that Dylan developed the PE at a later time.

73. It was felt that Dylan's symptoms at his original presentation to Rockingham Hospital, and later at FSH, could reasonably be explained solely by pulmonary oedema, and he then developed an embolism sometime during his hospital admission at FSH.[57] However, it could not be ruled out that Dylan had a pulmonary embolism from an early stage once in hospital. Although the tests, such as a chest x-ray and ECG done early in the piece at Rockingham Hospital, did not suggest a pulmonary embolus, it was noted by Dr Willis that they are not particularly sensitive tests for this condition.[58]

74. Further, it was not a case of 'either/or', as he could have been suffering from both conditions at the same time.[59]

75. Dr Ram Tampi is a Clinical Haematologist. Dr Tampi has given expert evidence in the Coroner's Court previously in relation to deaths caused by pulmonary embolisms, notably in the *Keogh* inquest referred to above. Dr Tampi reviewed Dylan's medical records in the context of Dylan's parents' letter to the Coroner, and provided an expert opinion to the Court on Dylan's medical care.

76. In Dr Tampi's opinion, when Dylan presented to FSH it was quite reasonable for his symptoms of persistent tachycardia and low oxygen saturations to be attributed to fluid overload and pulmonary oedema, in the context of other symptoms and his known nephrotic syndrome.[60] Dr Tampi agreed with Dr Willis that in the population in which Dylan fell as a patient, "it would be extremely rare for a pulmonary embolus to enter the equation."[61] Therefore, it was reasonable for the focus to be on the diagnosis of pulmonary oedema.

77. However, when Dylan's symptoms of tachycardia and low oxygen saturations persisted, Dr Tampi suggested pulmonary embolism would need to be considered, with the possibility that it was co-existent with pulmonary oedema.[62]

[55] T 20.
[56] Exhibit 1, Tab 6.
[57] T 20.
[58] T 19.
[59] T 20.
[60] T 31.
[61] T 32.
[62] T 32.

PROPHYLACTIC ANTI-COAGULATION

78. Anticoagulation is administered after a diagnosis of VTE or PE, but can also be considered for use prophylactically in patients considered to be at high risk of developing a clot. However, Dr Willis advised that the use of prophylactic anticoagulation is not generally recommended or practised in paediatric patients with steroid sensitive nephrotic syndrome. This is because of the low incidence of clinically significant thrombo-embolic events (such as PE) as against the relatively high risk of bleeding complications with the use of anticoagulation.[63]

79. Dr Willis explained in his evidence that doctors recognise that "the nephrotic state is prothrombotic,"[64] so they know there is a risk, but, as noted above, in the context of paediatric patients, it is not high on the list of risks and complications.[65] Dr Willis acknowledged that there is a gradation in age in terms of risks, so adolescents are at slightly higher risk than young children, but the risk is still significantly less than for adults with nephrotic syndrome. This was felt to apply to Dylan, even though he was the height and build of an adult male by this stage.[66]

80. Dr Willis cited a study published in 2013 that noted there are no randomised controlled trials demonstrating the safety and efficacy of pharmacologic thromboprophylaxis for the prevention of nephrotic syndrome-related VTE in either adults or children. Strong recommendations for or against pharmacological prophylaxis must therefore await definitive trials. In the meantime, non-pharmacological strategies to limit the likelihood of VTE in childhood nephrotic syndrome patients are suggested, such as regular ambulation, adequate hydration, avoidance of central venous catheter whenever possible and the use of graduated compression stockings and/or compression devices for bedridden children.[67]

81. With the specific exception of the rare condition Congenital Nephrotic Syndrome, Dr Willis advised that a prophylactic approach to anticoagulation is not recommended in paediatric nephrology and is not promoted in any paediatric guideline or recommendation that he reviewed, including from hospitals within Australia and internationally. The new PCH guideline, which at the time Dr Willis prepared his statement was yet to be endorsed, takes a similar position.

82. Dr Willis gave evidence that he has also spoken with his colleagues, both locally and elsewhere, about Dylan's case and the question of prophylactic anticoagulation, and all were in "universal agreement that if a similar case arose again, nobody would recommend prophylactic anticoagulation."[68] Dr Willis explained that a complication of anticoagulation is the risk of bleeding,

[63] Exhibit 1 Tab 6.
[64] T 13.
[65] T 13 – 14.
[66] T 15, 17.
[67] Exhibit 1, Tab 6B.
[68] T 21.

so that risk needs to be balanced against the possible benefit.[69] Dr Willis indicated that if Dylan had survived his pulmonary embolism, he may well have been a candidate for anticoagulation, as the level of risk had changed, but in the ordinary case the risk of bleeding would outweigh the likely utility.[70]

83. Dr Swaminathan is a Consultant Nephrologist and the Head of Nephrology at FSH. Dr Swaminathan was not involved in Dylan's medical care but was the Head of the Department when he was hospitalised.[71] Dr Swaminathan primarily treats adult patients, but also treats adolescent patients transitioning to adult care, such as Dylan. Dr Swaminathan reviewed Dylan's medical records and read some of the relevant statements, as well as discussing the case amongst her colleagues.[72] Dr Swaminathan agreed with Dr Willis' evidence that Dylan's care was a rare and unexpected event.[73]

84. Dr Swaminathan explained that in adult patients with nephrotic syndrome, the risk of thrombosis and thrombolytic events, such as pulmonary embolism, is higher than in paediatric patients, although it is still not common. There are particular patients who have a higher risk than others, depending on the type of nephrotic syndrome, with one high risk variety being glomerulonephritis, which is predominantly an adult disease. Nevertheless, even in those cases, it will be a clinical judgment in each case as to whether a patient will be put on prophylactic anticoagulation therapy, as it remains a balancing exercise between the risks of thrombosis against the risks of anticoagulation therapy.[74] Dr Swaminathan described it as a "grey area"[75] and explained that a doctor would consider several factors in reaching a decision.

85. Dr Swaminathan mentioned the risk factors would include how long the patient has had the disease, how severe it is, age and other co-morbidities, and any plans for a long plane journey or likelihood of being immobile for long periods. Previous history of clotting problems/risk factors in the past is also a significant consideration, and a past history of a blood clot will be an important factor in making the decision whether to commence therapy.[76]

86. In a case like Dylan's, being a paediatric patient with minimal change disease who has responded well to steroid treatment and has no past history of clotting, Dr Swaminathan expressed the view that she would expect they would be "on the lower end of almost not considering prophylactic anticoagulation,"[77] although she did not criticise the decision to administer Clexane.

87. Dr Tampi was asked whether in Dylan's case the administration of a therapeutic dose of Clexane, rather than prophylactic doses, could have

[69] T 21.
[70] T 22, 26 - 27.
[71] T 40.
[72] T 46 – 47.
[73] T 54.
[74] T 43.
[75] T 43.
[76] T 44
[77] T 45.

altered the outcome in his case. Dr Tampi responded that it was "really hard to say."[78] Dr Tampi did not think there was enough evidence to say that there was a pulmonary embolus at an earlier stage, so the prophylactic doses were appropriate at the time.[79] Dr Tampi noted the risk of bleeding, either gastrointestinal or even bleeding into the brain, can occur with larger doses, so without a strong suspicion that there is a pulmonary embolism, it was not appropriate to administer a larger dose.[80]

88. Dr Tampi commented that the "shifting nature of the problem meant that it was very difficult to know exactly when to approach thrombolytic therapy"[81] in Dylan's case. Dr Tampi explained that usually it would only be considered after a diagnosis of pulmonary embolism has been made; for example, in Dylan's case, the echocardiogram demonstrating a thrombus. A D-Dimer could also be performed and this could then lead to a CT pulmonary angiogram (CTPA). However, D-Dimer tests are not a strong indicator of pulmonary embolism in patients with nephrotic syndrome as the coagulation pathway is in an activated state resulting in an elevated D-Dimer level.[82] Therefore, Dr Tampi considered it appropriate that no such tests were ordered until there was a strong suggestion of a PE.[83]

89. In Dylan's case, Dr Tampi suggested that Dylan's deterioration may have been due to "disseminated intravascular coagulation, …. which is a pathological process characterised by activation of the clotting cascade that results in the formation of blood clots in the small blood vessels."[84] Dr Tampi explained that Dylan may have developed a small clot in his deep veins, which may have then triggered more clotting process to take place due to inflammation activating the coagulation system. The body has natural enzymes to dissolve the clot, but eventually this process can be overwhelmed by the clot propagation, leading to Dylan having numerous clots in his system.[85]

90. Dr Tampi indicated that anti-coagulant therapy would be the mainstay of treatment in such a case and, in the early stages, there would be a reasonable chance of success. However, as time progresses, the chances become less and less.[86]

91. It was explained at the inquest that the doses of Clexane administered to Dylan would have helped to prevent a clot from forming, but it was unlikely to be effective at treating an already established clot.[87]

[78] T 33.
[79] T 33.
[80] T 34.
[81] T 34.
[82] T 35.
[83] T 37.
[84] T 33.
[85] T 33.
[86] T 33.
[87] T 49.

TIMING OF DIAGNOSIS

92. Dr Tampi's comment above raises the question whether there should have been earlier investigations to make the PE diagnosis.[88]

93. Dr Willis, Dr Tampi and Dr Swaminathan all agreed that the approach of the doctors to Dylan's medical care at Rockingham Hospital and FSH was reasonable in circumstances.[89]

94. Dr Willis was asked whether, given the potentially fatal outcome of a pulmonary embolus, he considered greater emphasis should have been given to management of that risk, rather than pursuing the more obvious diagnosis of pulmonary oedema. Dr Willis noted that Dylan was given more than one dose of Clexane, which showed thrombosis was considered, but felt that the most likely diagnosis was properly pursued and treated. Dr Willis indicated that this was appropriate, "until such time as it became evident that there was something else going on."[90]

95. Similarly, Dr Swaminathan emphasised that in medicine, it is taught that "common things are common"[91] and it is important for doctors not to become distracted by rare possibilities. This is the approach taken by the doctors who treated Dylan at Rockingham Hospital and FSH, focussing on his obvious oedema and the statistically high likelihood his other symptoms of tachycardia and breathlessness and low oxygen saturations were a result of pulmonary oedema. The other possibilities that could arise based on the same symptoms, such as a cardiac event, pneumonia or pulmonary embolic event, were far less likely.[92] Dylan's age made a cardiac event unlikely and he showed no signs of infection. Therefore, pulmonary oedema was the more likely diagnosis, although it was recognised with the administration of Clexane that in the background was the potential for a thrombosis to develop.[93]

96. Dylan responded well to the intravenous frusemide, which was intended to get rid of the excess fluid, which was a further sign supporting the pulmonary oedema diagnosis.[94] An original chest x-ray at Rockingham Hospital showed signs of pulmonary oedema but a later one taken at FSH was reported as normal, with no evidence of fluid overload, therefore showing an improvement following the administration of frusemide and good urine output.[95]

97. However, when some of his symptoms persisted, further tests were conducted, including an echocardiogram that revealed the presence of the massive pulmonary embolism. Despite treatment, he did not survive. Dr

[88] T 85.
[89] T 37, 53.
[90] T 20.
[91] T 55.
[92] T 47 – 48, 59.
[93] T 49.
[94] T 49 - 50.
[95] T 51.

Swaminathan was uncertain whether an earlier diagnosis and treatment would have influenced the outcome.[96]

98. Dr Willis had no criticism to make of his colleagues, and felt they "considered all the things that [he] would have considered."[97] Dr Willis was confident in saying that his adult care colleagues in intensive care and nephrology and emergency at FSH were all better placed to diagnose and manage the pulmonary embolus than his colleagues who work in the PCH, where it is a "very, very rare presentation."[98] Dr Willis indicated that he has been practising in this area of medicine for over 30 years and he has never previously come across a child suffering a pulmonary embolism in the context of nephrotic syndrome.[99] In his view, while the outcome in Dylan's case was tragic, his care was appropriate and the investigations were appropriate. Dr Willis did not form the view that the pulmonary embolism should have been diagnosed at an earlier stage, based upon Dylan's presentation and history, but he accepted he was not an expert in this area and deferred to Dr Tampi.[100]

99. Dr Tampi was asked if his review suggested any changes that might be made for the future. Dr Tampi noted the information provided by FSH that in the event there is another case where there is sinus tachycardia, shortness of breath and general chest pain, a right ventricular echocardiogram would be recommended to help the medical team to decide whether there was a possibility of a pulmonary embolism. This is the test that identified the PE in Dylan's case, but sadly too late for treatment to be effective. Dr Tampi considered FSH's new policy to encourage doctors to do such a test at an earlier stage was a positive step towards diagnosing a similar event in another person.

LESSONS THAT MIGHT BE LEARNED FROM DYLAN'S DEATH

100. It is difficult to say with any certainty whether earlier diagnosis and treatment of his pulmonary embolism would have changed the outcome in this case, but it seems to be agreed that it would have given Dylan the best chance of survival. One of the difficulties is that it is unclear when Dylan developed the fatal clot. It was indicated that it may have been a sudden onset that occurred after he was transferred to the ICU, around the time of his deterioration on 1 August 2015, or it may have developed much earlier.[101]

101. Dylan's death highlights the importance of doctors keeping an open mind and constantly re-evaluating their diagnosis if symptoms such as tachycardia and breathlessness persist. Dr Swaminathan believes this is

[96] T 54.
[97] T 24.
[98] T 13.
[99] T 13.
[100] T 24 - 25.
[101] Exhibit 1, Tab 9 [43].

what the doctors did in this case, which ultimately led to the diagnosis of PE, when Dylan was not getting better.[102] Sadly, by that stage it was too late.

102. Dr Swaminathan indicated that Dylan's case will form part of her teaching and training of more junior doctors.[103] While it must be acknowledged that what happened to Dylan was unexpected and extremely rare, so the likelihood that it will occur again anytime soon at FSH is small, Dr Swaminathan said she emphasises with students the need to always keep in the back of their mind the worst that can occur, and to try to exclude any possibility that there might be something occurring that could kill a patient.[104] Nevertheless, Dr Swaminathan also spoke of the need to avoid over-treating patients, as the treatments can have their own complications, such as anti-coagulation. Therefore, it does not mean they will take a more pro-active approach to anti-coagulation in such patients, but the risk will remain to be assessed.[105]

103. Dr Willis had known Dylan and his family well, and he indicated in his evidence he had thought about Dylan many times and considered whether anything more could or should have been done. However, in retrospect, Dr Willis had concluded that he did not think he would have managed Dylan any differently, and if a similar case reoccurred, he did not think he or any of the other doctors would manage it any differently. He expressed the view that Dylan's death was "a tragic event resulting from a rare complication of a rare condition."[106] Despite giving the matter considerable thought, Dr Willis was unable to suggest any changes that might be put in place to avert a similar outcome.

104. I made it clear at the conclusion of the inquest that I would not be making any adverse comments or findings in relation to the medical care Dylan received. I accept the experts' opinions that Dylan's death was a tragic and rare event.[107]

105. As noted above, FSH reviewed its procedures and implemented a change following Dylan's death. Dr Paul Mark, the Director of Clinical Services at FSH, felt that bedside echocardiography could be used as a screening tool in a case like Dylan's, as it may identify right ventricular dilation and dysfunction, which can be associated with massive PE. This could then trigger the performance of confirmatory testing such as a CTPA, or if the patient is unstable like Dylan, then a move to the administration of a thrombolytic agent earlier.[108] Therefore, FSH ICU has implemented a separate on-call ICU echocardiography roster to ensure that such a study can be performed at an ICU patient's bedside at any time at the direction of an ICU Consultant.[109] As Dr Tampi agreed, this is a positive step.

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[102] T 56.
[103] T 54.
[104] T 54.
[105] T 55, 57.
[106] T 26.
[107] T 60.
[108] Exhibit 1, Tab 5.
[109] Exhibit 1, Tab 5.

CONCLUSION

106. Dylan was a young man verging on adulthood who had a history of nephrotic syndrome that was well-managed. It was hoped his condition might resolve on its own, which I am told it often does, but if it did not then there were plans in place for his care to be transitioned from paediatrics to adult care at FSH. There was nothing to raise any concerns about his prognosis and prospects at that time.

107. Sadly, he had a further relapse and developed a rare and unusual complication of a massive pulmonary embolism. He was in the ICU at FSH when the pulmonary embolism was discovered. All appropriate steps were taken to try to treat it immediately, but it could not be dissolved in time to save his life and he died in hospital on 1 August 2015.

108. Dylan's parents requested a full coronial inquiry to ensure that any changes that might be made to save another child are made. The doctors involved in Dylan's medical care, and the management at FSH, all cooperated fully with the coronial inquiry. However, what the investigation has revealed is that Dylan's death was a rare and unexpected event that was difficult to predict. What changes that might be made to diagnose a similar case sooner have been made, and hopefully that will give Dylan's parents some small solace. Further, they can be satisfied that a full and thorough investigation into his death has been conducted, so that they know that this was not a case where there was a failure in the medical treatment that might otherwise have saved his life.

S H Linton
Coroner
23 September 2019