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CONTENTS

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Next Edition: December 2016

EDITORIAL

In our third edition for the year, we explore three very sad cases involving young children who all had relatively uncommon conditions. The healthcare-related death of a child is always a profoundly upsetting experience for everyone concerned – from the treating clinicians, to those called upon to investigate the circumstances of the death, and most of all, to the parents and relatives who suffer such a loss. There are usually many difficult questions that need to be asked and answered. What is all too jarring is the death of a previously well child, who had no known natural diseases or medical conditions, and who had no real reason to become unwell. In those scenarios, the relatively rapid course that can progress from a child appearing completely well to being gravely ill is particularly distressing. Compounded by the inability of young children to understand or communicate their symptoms, parents and clinicians are left wondering what could have, or should have, been done sooner.

The paediatric stories in this edition have been undeniably tough to write, and will be confronting to read. They are stories that we may remember for a long time to come. Yet in many ways, this is not an edition about paediatric illnesses. It is about seeing the signs, even if you do not see the diagnosis. The cases presented here serve to highlight the universal themes that come up time and again, in the care of adults as well as children. In each of the cases, the parents sought medical attention on multiple occasions, however, the treating staff were arguably blinkered to their concerns by preformed assumptions, and did not react to a few vital cues that could lead them to recognise a sick patient and investigate further. If we remember these stories we can change our practice for the better.

The expert commentary for this issue has been written by Dr Annie Moulden, a paediatrician with extensive experience in quality and safety networks. Her commentary explores the importance of a framework that every clinician should have for safely managing the undifferentiated patient. When a person is suffering from a rare condition (one that we may never have come across before), and the diagnosis eludes us, there are ways in which we should approach the situation, irrespective of the missing pieces of the puzzle. Being cognisant of the physiological changes that are occuring, even without a clear picture of the prevailing disease process, allows for specific measures to be put in place to manage acute complications and possibly avoid a catastrophic event. The trigger points do not need to be complex. Sick or not sick, admit to hospital or not. Focus on the warning signs – pain out of keeping with appearance, repeat presentations, family concerns, abnormal vital signs – and effect a plan that is simply and safe. Adjustments and fine tuning can follow later.

In this issue, we are very fortunate to welcome back Dr Fennessy as a case author, and also to welcome a new member to our team of authors, Dr D'Costa, an intensivist and the State Medical Director of DonateLife. Both authors share their sage insights with us on the lessons learned from these tragic deaths.

Our new website is finally here and we are eternally grateful to Mr Alexander Gillard who designed and built the new site. He has given us a fresh new look with a more intuitive and functional layout. Please take a look at our features, for easy access to our resources and editions, and for a sneak peek at our latest exciting venture which is coming soon.

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ACKNOWLEDGEMENTS

This initiative has been made possible by collaboration with the Department of Forensic Medicine (DFM), Monash University, Victorian Institute of Forensic Medicine (VIFM) and Victorian Managed Insurance Authority (VMIA).

FEEDBACK

The editorial team is keen to receive feedback about this communication especially in relation to changes in clinical practice. Please email your comments, questions and suggestions to: clinical.communique@vifm.org

CASE #1 INTRACTABLE PAIN, TRAGIC DEATH

Case Number:
COR 4449/07(0) QLD

Case Précis Author:
Dr Rohit L D'Costa
MBBS, FRACP, FCICM

CLINICAL SUMMARY

RS was a 2 year old boy who was taken by his mother to their family GP in the context of general unwellness and cervical lymphadenopathy. A presumptive diagnosis of mumps was made and the GP advised symptomatic treatment. Over the next three days however, RS developed worsening malaise, anorexia and abdominal pain, and was taken by ambulance to the local hospital where doctors suspected intra-abdominal pathology, possibly intussusception. Investigations including x-rays and blood tests were performed which did not reveal significant abnormalities.

Arrangements were made to transfer RS via helicopter to the regional base hospital with paediatric services. On arrival at the base hospital, RS was noted to be febrile (38.7°C), tachycardic (162 bpm), tachypnoeic (28 breaths/min), and in pain. He was given opioid analgesia in the emergency department prior to transfer to the paediatric ward.

On the following morning ward round, the paediatrician elected to continue the analgesic regimen and did not agree with suggestions made by the resident doctor to add morphine or to perform further septic workup or commence empiric antibiotics and intravenous fluids.

Intussusception was now considered unlikely on the basis of the clinical picture and his age, but signs of meningism (Brudzinski's sign and Kernig's sign)* were found by the paediatric team, prompting the performance of a lumbar puncture (LP). Clear CSF drained. No blood cultures were drawn as the consultant paediatrician's impression was that the child was suffering from a viral illness.

Into the night after transfer, RS had ongoing irritability and inconsolable pain which was not relieved by the simple non-opioid analgesia that had been prescribed.

On the following morning ward round, the paediatrician elected to continue the analgesic regimen and did not agree with suggestions made by the resident doctor to add morphine or to perform further septic workup or commence empiric antibiotics and intravenous fluids. He did, however, organise further imaging of the abdomen, which did not yield a diagnosis.

A surgical review was also sought, and on reviewing RS later that evening, the surgical team felt an exploratory laparotomy was warranted.

A period of several hours passed with RS becoming increasingly distressed before an intravenous cannula was inserted and blood was drawn for cultures, FBE, electrolytes and CRP.

It was the abnormalities in these results, and in particular the markedly elevated CRP result of 444mg/L (normal range up to 5mg/L), that prompted further consideration of bacterial infection. A decision was made in consultation with a second paediatrician (who was to be on-call that evening) to commence broad spectrum antibiotics. A surgical review was also sought, and on reviewing RS later that evening, the surgical team felt an exploratory laparotomy was warranted. It was during the pre-operative anaesthetic review that the duty anaesthetist recognised RS's grave condition, and the need for urgent transfer to a tertiary referral hospital. A retrieval team was mobilised and on their arrival that night, a decision was made to intubate RS for transfer in view of his extremely ill state. RS suffered a cardiac arrest on intubation and was not able to be resuscitated.

PATHOLOGY

Autopsy revealed the presence of a small intestinal volvulus without associated features of infarction. The pathologist noted this was not demonstrated in life by radiology and may have been an agonal event. There was severe bronchopneumonia and cervical and abdominal lymphadenopathy. Blood culture results from the sample drawn prior to death grew group A streptococcus.

CASE #1 INTRACTABLE PAIN, TRAGIC DEATH (Continued)

INVESTIGATION

An inquest was held following the release of the report of an investigation by the Health Quality and Complaints Commission (HQCC). Amongst other matters, the inquest sought to confirm the cause and circumstances surrounding RS's death, and the adequacy and appropriateness of his medical treatment at the various stages of his care. The expert and witness statements collated in the HQCC report formed the basis of the information considered at inquest.

Expert opinions were provided by an infectious diseases physician and a general practitioner who were asked to comment on the pre-hospital care. Both considered the initial diagnosis of mumps unlikely as RS had been fully vaccinated, but thought that the treating GP's examination and management plan were reasonable. Independent experts also examined the records and referral letter from the local hospital, and it was confirmed that copies had been sent with RS to the regional base hospital.

The Coroner found that RS died from Streptococcal Toxic Shock following a delay in recognition and treatment of severe infection.

The treating paediatrician at the regional base hospital told the inquest he had relied upon the referral letter from the local hospital and the handover by his resident of RS's condition, when he attended to assist with the lumbar puncture. He conceded he had not read the medical charts and had not been aware of RS's vital signs or the prescription of opioid medication on RS's arrival at the ED. It was noted that on the day of his death, RS had not been physically reviewed by a consultant between the morning paediatric ward round and the evening surgical review.

RS's parents recalled watching their son scream in excruciating pain through the night after the LP. They had requested stronger pain relief and expected further medical reviews and investigations but none occurred overnight. Testimony from nursing staff described repeated requests to medical staff to chart morphine as RS was "extremely irritable…with arched posturing", "moaning", and "wailing" with pain all over.

An expert opined that the failure to perform a septic work-up in a child who was sick enough to warrant a lumbar puncture was a "significant omission".

With toxic shock due to group A streptococcus, the initial presentation can mimic a range of infectious and non-infectious conditions, with pain a key feature.

Furthermore, the first dose of antibiotics was not administered until 24 hours after RS arrived at the regional base hospital.

CORONER'S FINDINGS

The coroner found that RS died from Streptococcal Toxic Shock following a delay in recognition and treatment of severe infection. He was satisfied with the treatment provided by the GP and the local hospital but was critical of a number of aspects of RS's care at the regional base hospital as directed by the paediatrician, namely:

- The incomplete history and inquiry into the observations and treatments given to RS prior to admission to the ward, which meant that the fever and severity of the pain were overlooked.
- The initial reluctance to consider alternative serious diagnoses after bacterial meningitis was ruled out on lumbar puncture, which led to a critical delay in septic workup.
- The reluctance to supplement simple analgesia with opioids despite repeated concerns expressed by RS's parents, the nurses, and junior medical staff.
- The lack of consultant review of the patient between the morning ward round and the pre-terminal events.

The coroner referred to the recommendations already made by the HQCC and their subsequent implementation by the health service. They included the introduction of the Children's Early Warning Tool (CEWT), formal processes for senior medical staff handover, and improved education on septic shock in children.

AUTHOR'S COMMENTS

This case illustrates the challenges that may arise in the diagnosis and management of an undifferentiated acute illness. With toxic shock due to group A streptococcus, the initial presentation can mimic a range of infectious and non-infectious conditions, with pain a key feature. Septic shock and associated multiple organ failure eventually ensue. Early antibiotic administration is crucial – this needs to be empiric as blood cultures may reveal the diagnosis too late.

The findings of the coroner are salutary. There is a clear imperative for a receiving doctor to gather and consider all information available regarding a transferred patient, especially when faced with diagnostic uncertainty. They need to consider a broad differential in these cases and not be anchored to a particular diagnosis or be hampered by confirmation bias. Finally, it is important to fully consider the causes of, and adequately treat pain. The child's parents felt that RS's pain was at times ignored. It may well have been the closest feature to a cardinal symptom in this case.

RESOURCES

Stevens DL. Streptococcal Toxic-Shock Syndrome: Spectrum of Disease, Pathogenesis, and New Concepts in Treatment. *Emerg Infect Dis* [Internet Journal]. 1995. Available from: http://wwwnc.cdc.gov/eid/article/1/3/95-0301.

KEYWORDS

Toxic shock syndrome, group A streptococcus, septic shock, pain, paediatric, missed diagnosis

** Brudzinski's sign – Flexing the patient's neck causes flexion of the hips and knees.*

Kernig's sign – Flexing the patient's hip 90 degrees then extending the patient's knee causes pain. Both signs are thought to be due to irritation of motor nerve roots passing through inflamed meninges as the roots are brought under tension. In this case, it was later suggested that RS stiffened in pain with any light physical contact, which had been interpreted as signs of meningism.

CASE #2 A DIPSTICK DILEMMA

Case Number:
2010/2935 QLD
Case Précis Author:
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B.Med, MForensMed
FFCFM (RCPA), FACEM

CLINICAL SUMMARY

TO was a 4 year old boy who in the 12 months leading up to his death had been experiencing sporadic episodes of discoloured urine, sleepiness and muscle pains.

TO's parents brought him to a family clinic for review on a number of occasions and reported their concerns which included his gait when walking up hills, back pain, sore legs, fevers, night time restlessness, and brief moments of dark, smelly urine. Each time TO was seen, his physical examination was normal. Over the course of his presentations, he was seen by four GPs and a paediatrician, and investigated with x-rays of his lumbosacral spine and hips, liver and renal ultrasounds, and urine biochemistry to check for calcium excretion.

The investigations were all normal. A urine microscopy sample showed protein ++ and blood +++, but no red or white cells, and was reported as "probable haemolysis of red blood cells". This test was repeated twice more with the same findings. Blood results showed elevated liver enzyme levels, but were otherwise normal, and a repeat sample a month later returned normal enzyme levels. A viral illness was thought to be the most likely diagnosis, and although a metabolic disorder was considered, it was not pursued as TO appeared healthy.

On one occasion, TO complained of back pain and difficulty passing urine. He had a temperature of 38.1°C and sensitivity to light. He was seen by another GP and referred to hospital. TO was assessed by an intern in the ED who noted dark urine and an otherwise normal examination. A dipstick urinalysis revealed a large amount of blood and protein.

The intern discussed her findings with her supervisor and as TO appeared clinically well, a diagnosis of viral illness was made and he was discharged home to be followed up by his GP for "resolution of haematuria". The hospital urine microscopy and culture results were later reported as normal and sent out to the GP who had not received a discharge letter and was unaware of the abnormal dipstick results.

Approximately two months later, TO woke complaining of knee pain which then extended to other parts of his body. His parents called an ambulance when he became increasingly lethargic. Ambulance crew arrived to find a pale, unwell looking child who had a Glasgow Coma Scale score of 10, a low temperature of 33.9°C and a fast pulse of 143 bpm. He was transported to hospital where he arrested 30 minutes after arrival. He was intubated and treated for a severe metabolic acidosis with hyperkalaemia. Despite aggressive resuscitation efforts, he died shortly afterwards.

PATHOLOGY

The cause of death at autopsy was severe rhabdomyolysis, and following specialist testing in France, the abnormalities were found to result from an LPIN1 gene mutation.

INVESTIGATION

Internal reviews were conducted by the hospital and the ambulance service. The hospital showed that it was implementing procedures to consolidate discharge processes in the ED and increase the ED's capacity and preparedness for emergency paediatric cases, as well as to improve alerts, communication and follow-up regarding test results. The ambulance service considered there was no deviation from clinical practice and standards.

It was felt that the actions of the GPs, and the care provided at both hospital visits was appropriate and conducted according to relevant professional standards.

TO's parents lodged a complaint with the Health Quality and Complaints Commission who conducted an investigation and provided their information to the Coroner's Office.

The parents expressed concerns over the actions of the GPs and paediatrician who had been consulted about their son, and the hospital staff who had been involved in his care at both hospital presentations.

The coroner identified a number of issues to explore at inquest, amongst them, whether there were earlier clinical signs which should have alerted medical staff to undertake further reviews and investigations.

Expert opinions were obtained from an emergency physician, a paediatrician, and a GP.

One expert highlighted that five urine samples had been sent in the months prior to TO's death, and everyone failed to recognise the pattern that the 'blood' on a 'dipstick' was not red or white cells on microscopy. Two of the experts believed that the description of the dark urine with the microscopy findings should have raised suspicion on subsequent presentations, however, the associated symptoms were not classical for rhabdomyolysis.

All the experts reflected that they had never seen a child present with spontaneous rhabdomyolysis. It was felt that the actions of the GPs, and the care provided at both hospital visits was appropriate and conducted according to relevant professional standards. They were uncertain as to whether the information from the discharge summary, if known, would have prompted further investigations by the GP that may have changed the likely diagnosis.

CORONER'S FINDINGS

The coroner felt there were missed opportunities to make the diagnosis of rhabdomyolysis and put in place a management plan that could have altered the outcome, but he was not critical of the doctors involved in TO's care for being unaware of the LPIN1 gene mutation.

The coroner remarked that *"any capacity to make a diagnosis of rhabdomyolysis was compounded by TO having a virtually unknown underlying condition and most of the time displaying atypical symptoms"*.

The coroner concluded by acknowledging the efforts of TO's parents for doing all they could for their son, and for advocating on his behalf since his death, to bring his rare condition to the attention of the medical world in Australia for future patients.

RESOURCES

Meijer I, et al. LPIN1 deficiency with severe recurrent rhabdomyolysis and persistent elevation of creatine kinase levels due to chromosome 2 maternal isodisomy. *Molecular Genetics and Metabolism Reports* 2015; 5: 85–88.
Available at:
http://www.sciencedirect.com/science/article/pii/S2214426915300458

KEYWORDS

LPIN1 deficiency, paediatric, rhabdomyolysis, missed diagnosis, genetic mutation

For a description on LPIN1 deficiency please refer to page 6.

CASE #3 PARENTAL GUIDANCE RECOMMENDED

Case Number:
47/2014 WA
Case Précis Author:
Dr Gerard Fennessy
MBChB, PGDipCEM, FRNZCUC, FCICM

CLINICAL SUMMARY

CT was a 2-year-old male who was otherwise well. His parents were both health professionals. He developed a high fever that persisted over two days, with decreased appetite and his parents noticed a lump in his neck. He was taken to a major teaching hospital ED where he was found to be alert but miserable, with dry lips, moist mouth and a sore throat. He was tachycardic, febrile, tachypnoeic and mildly hypoxic.

He had improved when seen by the paediatric registrar, Dr L. All his observations had returned to normal and he was felt to have an upper respiratory infection with cervical lymphadenopathy. He was given antibiotics, analgesia, oral fluids, and Dr L advised the parents to return if they were concerned. No blood tests were taken although the parents did question this.

CT continued to be unwell, with vomiting, diarrhoea, fever and reduced oral intake, developing a rash over his face and body. He was seen by his GP, Dr T, 2 days later. Dr T reviewed the ED discharge letter and felt the rash may have been due to the medications, so they were ceased.

CT's fever remained and the next day his parents took him back to ED. He was triaged as category 3, and seen within 30 minutes by Dr P, who was an experienced paediatric registrar.

CT was alert but miserable with dry cracked lips, sore throat, widespread blanching rash and conjunctivitis. He had a fever (39.5°C), heart rate of 160-177 bpm, respiratory rate of 44/min, and oxygen saturations of 97%. It was noted that the clinical picture was inconsistent with the parental reports, particularly of continued vomiting and lethargy. Because CT had represented, his case was further discussed with the paediatric consultant, Dr M, who also examined CT and agreed with a diagnosis of viral illness. The parents were concerned about the diagnosis and believed CT needed to be admitted. After further discussion with Dr P, it was felt he was able to be cared for at home, particularly as he had well-informed parents.

The following day, CT was reviewed by his GP again. He had been unwell for six days with no improvement. Dr T had access to the discharge letter and he felt the presentation was likely to be a viral illness.

Over the next day CT became more lethargic, had worsening diarrhoea, severe cough, and complained of increasing generalised pain. He also developed swelling of his feet, face and ankles.

That evening, CT was heard to be gurgling and was found unresponsive in bed by his parents who commenced CPR immediately. An ambulance was called and CPR was continued enroute, however on arrival at the hospital it was clear resuscitation attempts were futile. CT was asystolic and mottled with a pH of 6.8, a lactate of 15, and had not had cardiac output for over 40 minutes.

PATHOLOGY

Post mortem showed a soft dilated heart, pericardial effusion, pulmonary oedema and pneumonia with enlarged lymph nodes in the neck and around the airway. There was diffuse myocarditis and mild hepatitis on histopathology. Cause of death was given by the pathologist as myocarditis that was more consistent with originating from Kawasaki Disease (KD) than from a systemic viral illness.

INVESTIGATION

At inquest, the coroner heard that KD is a clinical diagnosis of an acute febrile vasculitis. It can mimic many other viral-like illnesses, and as there are no confirmatory tests available, requires a high degree of suspicion. It is usually self-limiting, but may respond to immunoglobulin or aspirin. Furthermore, myocarditis is a rare complication of KD and can precipitate fatal arrhythmias.

Although it was not documented, Dr P stated to the court that KD was considered, however Dr P did not believe this was consistent with the clinical findings. Dr P had coincidentally previously published a case series on KD. In discussing the diagnosis of viral illness, Dr M explained that the ED had seen many patients with similar symptoms over the preceding days.

CT's death was reviewed by two general paediatricians and an emergency paediatrician, and there was significant disagreement as to whether the diagnosis should have been clear to the treating doctors. Whether admission was warranted, as the parents had asked for, was also in dispute.

CT's parents gave evidence that they did not recall being asked to return to hospital if they had concerns. In fact, it appeared that they felt dissuaded from returning if there was no improvement.

It was agreed that three days prior to his death, CT had all the clinical signs of KD and this should have been considered (fever of at least five days duration; bilateral non-exudative conjunctivitis; erythema of the lips/oral mucosa; changes in the extremities; rash; and cervical lymphadenopathy). One reviewer stated however, that although KD was a reasonable diagnosis, a viral illness was more likely, and the doctors had to accept the uncertainty associated with this. It remained unclear whether admission and treatment with immunoglobulin or aspirin would have altered the course of the disease and ultimately prevented death.

CORONER'S FINDINGS

The coroner found that CT suffered a cardiac arrhythmia as a result of myocarditis, a consequence of his being in the acute phase of KD, and death arose by way of natural causes.

The coroner commented on the importance of remembering that distressed parents are vulnerable to receiving the wrong messages from clinicians.

There were no clear systemic issues identified as needing to be addressed by the health service. There were adequate strategies already in place, including senior clinician review for representations and written discharge information. It was thought that these could not be significantly improved upon to prevent a similar death.

However, importantly, it became clear during the inquest that the parents perceived they were unwelcome to return to hospital after the second review. A number of issues may have contributed to this, including the lack of blood tests, the diagnosis of viral illness and the refusal of a direct request from the parents for admission. The coroner commented on the importance of remembering that distressed parents are vulnerable to receiving the wrong messages from clinicians. Although admission may not have altered the outcome for CT, it may have led to better hydration and recognition of deterioration, and also may have assisted the parents to accept the outcome.

CASE #3 PARENTAL GUIDANCE RECOMMENDED (Continued)

This case is a reminder that we should always remain open to alternative diagnoses, particularly when the provisional diagnosis (viral illness) is one of exclusion. CT was seen by several experienced health professionals prior to his death, including one who had seen several cases of KD.

It is not clear whether the death could have been prevented, even if the diagnosis of KD was made and treatment initiated. Nonetheless, the coroner highlighted some important issues regarding the role that parents play as advocates for their children, notably that on a number of occasions CT's parents expressed significant concerns about their child and requested admission because they 'knew' something was wrong. It may seem unfathomable that the parents felt unwelcome to return to hospital – yet that was their experience.

RESOURCES

Logue E, et al. Characteristics of patients and families who make early return visits to the pediatric emergency department. *Open Access Emerg Med* 2013; 5: 9-15.

Robinson K, Lam B. Early emergency department representations. *Emerg Med Australas* 2013; 25(2): 140–146.

KEYWORDS

Kawasaki disease, representation, paediatric, missed diagnosis, myocarditis

EXPERT COMMENTARY

LISTEN TO PARENTS AND LOOK AT THE VITALS

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Child and Adolescent Sub-Committee member, Consultative Council Obstetric and Paediatric Morbidity and Mortality

The paediatric cases in this edition highlight the difficulties faced sometimes in recognising the seriously unwell child, especially when the underlying diagnosis may not be readily apparent. Whilst earlier recognition and intervention may not have changed the outcomes for these children and their families, the lack of appreciation by clinical staff of how unwell these children were, is something we must reflect on.

Hundreds of thousands of children present every day across Australia to healthcare facilities, the vast majority with self-limiting or relatively benign conditions. Clinicians need a framework to ensure they do not miss recognising the small number of very unwell children.

Every State in Australia now has Paediatric Observation and Response charts to assist clinicians in determining whether observations for a particular age group fit within reasonable limits, and to outline escalation processes where they do not.

The recognition of unwell children poses some unique challenges for clinicians, not the least of which is knowing what is normal and what is not, but children also bring with them a major asset – parents as experts on their child's 'normal state'. Parents know their children intimately, especially when they are unwell. We need to listen to their concerns carefully.

Severe pain and distress in a young child is unusual – waking in pain and complaining of 'all over body pain' should concern us. Similarly, reluctance to eat or drink for 'days on end'.

The child with Kawasaki disease presented to both his GP and tertiary hospital ED on multiple occasions. On the second ED presentation, he had a number of clinical features of KD, however he was discharged home with a diagnosis of a viral illness. It is important we reflect on his parents' statement to the coroner that they perceived they were not welcome to return to the ED after that attendance.

Toxic shock syndrome from Group A Streptococcal infection is again, a relatively uncommon diagnosis however this young patient's parents were very clear that all was not well, particularly with regard to his level of pain and distress. He also had a persistent tachycardia. Whilst this is often assumed to be due to fever, distress or both, when the tachycardia persists it can be critical in identifying the seriously unwell child.

The third case involves a rare genetic defect causing rhabdomyolysis and whilst many clinicians would not have made that diagnosis, the presentation of migrating body pain with discoloured urine is sufficiently unusual to have moved him into the 'of concern' category.

Every State in Australia now has Paediatric Observation and Response charts to assist clinicians in determining whether observations for a particular age group fit within reasonable limits, and to outline escalation processes where they do not.

Listening to parents' descriptions of their child's symptoms and behaviour, and carefully considering the vital signs, are the two critical factors in the early identification of the unwell child.

LPIN1 DEFICIENCY AND RHABDOMYOLYSIS

Rhabdomyolysis is a breakdown of skeletal muscle, with release of myoglobin into the bloodstream which can cause damage to the kidney cells. Symptoms include: dark urine, decreased urine production, muscle stiffness, pain and fatigue. There are multiple causes of rhabdomyolysis including metabolic and genetic factors. LPIN1 deficiency is a genetic abnormality causing skeletal muscle to breakdown and has been associated with sudden death in young children. It is an extremely rare disease, which up until TO's death probably had never been described in Australia. The clinical features of LPIN1 include: life-threatening rhabdomyolysis and acute metabolic decompensation; normal strength, development and neurological examination between attacks; and normal fat distribution. Although rhabdomyolysis in LPIN1 may occur spontaneously, triggers include: fasting, fever, exercise, dehydration, overheating, and intercurrent illness.