



HEALTHCARE SAFETY  
INVESTIGATION BRANCH

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# INTERIM BULLETIN

## UNDIAGNOSED CARDIOMYOPATHY IN A YOUNG PERSON WITH AUTISM

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This interim bulletin contains facts which have been determined up to the time of issue. It is published to inform the NHS and the public of the general circumstances of events and incidents and should be regarded as tentative and subject to alteration and correction if additional evidence becomes available.



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## NOTIFICATION OF EVENT AND DECISION TO INVESTIGATE

HSIB was notified by a district general hospital of the death of a young patient who had undergone an MRI scan under general anaesthetic to investigate their recurrent headaches. The patient was being treated for growth hormone deficiency and had autism spectrum disorder and an associated learning difficulty. During the scan, the patient suffered unexpected deterioration, which was subsequently discovered to have been caused by an undetected heart condition (cardiomyopathy<sup>1</sup>). The patient was stabilised in the hospital intensive care unit and subsequently transferred to the Paediatric Intensive Care Unit at a regional children's hospital but died three days later.

Undergoing general anaesthetic to facilitate MRI scanning is well established in the UK. The Royal College of Anaesthetists (RCoA) have published guidance to minimise the risks associated with carrying out anaesthetics away from the theatre environment. Deaths under general anaesthetic is a rare event, and the RCoA publishes information on the risk of death associated with having an anaesthetic which states that for “...[for] patient[s] having non-emergency surgery... death is very rare. An exact figure is not known, but it is around 1 death per 100,000 general anaesthetics” and that “...the risk of a child dying from a general anaesthetic is around 1 in 40,000. However, if the child is healthy and having non-emergency surgery, the risk is much less, probably less than 1 in 100,000”<sup>2</sup>

The scoping investigation conducted by HSIB suggests there are opportunities to improve the way patients are assessed for suitability for anaesthetics. It also looked at how autism, learning difficulties or learning disabilities have an impact when patients are accessing healthcare services.

<sup>1</sup> Cardiomyopathy is where the heart becomes enlarged and reduces its ability to pump blood effectively.

<sup>2</sup> Risks associated with your anaesthetic Section 15: Death or brain damage. <https://www.rcoa.ac.uk/system/files/15-DeathBrainDamage2017.pdf>



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The reference event highlighted potential national safety risks linked to care pathways, human factors, awareness and training regarding patients with autism, learning difficulties and learning disabilities, consent, and clinical practice guidance when undertaking MRI scans under anaesthetic.

Following the preliminary investigation, the Chief Investigator authorised a full investigation as the risk met the following criteria:

**Outcome Impact - what was, or is, the impact of the safety issue on people and services across the healthcare system?**

- Undetected cardiomyopathy can cause sudden unexpected death arising from a variety of different activities which can put additional strain on the heart. Collapse due to cardiomyopathy may also occur spontaneously.
- Where such incidents occur in healthcare settings, this can undermine patients' confidence and trust in healthcare services.

**Systemic Risk - How widespread and how common a safety issue is this across the healthcare system?**

- Pre-anaesthetic assessment is intended to determine patients' suitability for anaesthetic and prevent on-day cancellation of procedures. There is a risk that existing guidance does not sufficiently detail the investigations and examinations that should be undertaken, particularly with children and people with additional needs.
- The sudden death of a child during a routine procedure under anaesthetic is rare. However, it is more common for children with autism and/or learning disabilities/difficulties to require medical imaging under general anaesthesia or sedation. As in the reference event, a small number of these children may have diseases that, if undetected, could lead to an adverse outcome.



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## **Learning Potential – What is the potential for an HSIB investigation to lead to positive changes and improvements to patient safety across the healthcare system?**

- There is limited guidance on the content of pre-anaesthetic assessments, including the recording of physical observations and examinations.
- There is a lack of training and awareness within mainstream healthcare services on how to optimise care for patients with autism, learning difficulties and learning disabilities.
- The guidance on seeking consent for patients undergoing non-urgent anaesthetics for diagnostic procedures is limited and the opportunity exists to review these arrangements.
- There is no published guidance on the threshold for terminating imaging procedures under general anaesthetic should a patient's condition change in an unanticipated way. Similarly, there is limited guidance on the practice of remaining at the patient's side while undergoing MRI under GA. The guidance published by the RCoA suggests that:

*“Children presenting for anaesthesia outside the operating room may present challenges relating to the procedure, the environment, or physical, physiological and psychological challenges. Children may often require repeat treatments or investigations.”*

## **HEALTHCARE DELIVERY GOAL**

To promote the safety of general anaesthesia for children and people with autism, learning difficulties and learning disabilities undergoing diagnostic procedures by optimising pre-assessment, consent processes and further improving anaesthetic practice.



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## HISTORY OF THE EVENT

The patient was referred to her local hospital for an MRI scan under general anaesthetic to investigate recurrent headaches. The patient had been treated for the past ten years for growth hormone deficiency. She also had autism spectrum disorder and a learning difficulty.

The patient had severe anxiety about medical procedures and became agitated whenever she presented to health services. She attended a pre-assessment clinic with her mother, just over two weeks prior to the scan date, and was found to be fit for the anaesthetic.

At 07:30 hours on the day of the scan, the patient was admitted onto the ward by a member of the nursing team and a single set of observations were recorded. These were found to be abnormal (elevated heart rate and blood pressure) and this was escalated to the anaesthetist via the nurse in charge. These changes were attributed to her distress.

Shortly after the scan commenced at around 11:30 hours, a slow heart rate (bradycardia) of 37 beats per minute was detected and this was corrected using the drug atropine. The scan recommenced but a short time later was suspended again when the patient's heart rate became fast (tachycardia) at 140 beats per minute. This was also corrected but reoccurred, and after a third episode of tachycardia which was also corrected, it was agreed by the anaesthetic team to complete the scan to avoid a further, distressing visit to hospital and having another anaesthetic. The patient continued to be monitored during the remainder of the scan.

On completion of the scan, when the patient was physically assessed, it was noted that they had deteriorated significantly. Assistance was sought to help stabilise the patient. A cardiac ultrasound (echocardiograph) was undertaken which showed a grossly enlarged heart.



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A blood test was taken which confirmed that the patient was gravely unwell.

The patient was referred to the regional children's hospital who dispatched a retrieval team to transfer her for specialist treatment. Over the coming days the patient received intensive care but died on day four following the scan.

After the patient's death, a cardiac muscle biopsy was taken which was analysed at a specialist national laboratory. The results showed a mitochondrial disorder (Respiratory Chain Enzyme complex I deficiency). The post-mortem examination report confirmed cardiomyopathy and concluded that the cause of death was due to organ failure which occurred as a result of the decompensation of the patient's hearts function while under anaesthetic.

## **NATIONAL CONTEXT**

Hospitals with the facility to conduct MRI scans under general anaesthetic for children undertake between 1.5 and 7 scans per week, depending on the size and catchment of the Trust. The approach to pre-assessment may differ between hospitals, and the provision of additional support for patients with special needs may also vary.

The detection of diseases in patients with autism, a learning difficulty and/or a learning disability is challenging. In the context of an elective anaesthetic, the investigation will seek to establish the extent of the safety risks associated with patients with special needs highlighted in the reference event.

The national investigation will also examine the processes relating to preparing paediatric patients for elective anaesthesia. This will include the obtaining of consent - specifically considering how patients and families are given information regarding the risks and benefits of the procedure they are going to have; pre-assessment and understanding how published guidance is interpreted and operationalised.



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The investigation will work with stakeholders to develop safety recommendations to support clinicians to effectively communicate with, and assess, patients to optimise the chances of detecting diseases, some of which may be more prevalent in those with autism or learning disabilities.

## IDENTIFIED SAFETY ISSUES

The HSIB has analysed the reference event and has identified potential safety risks that will be explored within the national investigation.

Hospital staff may not be trained to recognise and manage the challenges associated with patients with autism, learning disabilities or learning difficulties. The patient was not seen by a learning disability specialist during their care, and their altered physiological markers were attributed to emotional distress.

Pre-anaesthetic assessment for non-operative anaesthetics in children does not include the routine recording of physiological parameters, and the published national guidance mandates local providers to develop their own processes rather than follow a standardised national process.

The MRI imaging environment has known risks for patients under anaesthetic, but these were mitigated in the reference event based on current national guidance. There is limited national guidance on the threshold for terminating non-operative procedures under general anaesthetic in the event of patients becoming unstable, and/or the practice of remaining in the scanning room with certain types of patient.

The investigation has also identified an opportunity to analyse the methods by which consent for anaesthetics is sought, and which provides detailed information for patients and their families on the risks and benefits



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of each aspect of their care; the potential choices they have, and the importance of having time to undertake shared decision making.

The investigation will therefore focus on;

- The current evidence base and guidance for anaesthetic pre-assessment clinics, on-day procedures which involve anaesthetics, consent in children, and the considerations for patients with special needs or who require reasonable adjustments to be made.
- The impact that autism, learning disabilities and learning difficulties have on mainstream healthcare service provision in relation to the safety risks identified, and the detection of diseases in patients with special needs.

## **NEXT STEPS**

The HSIB investigation will continue to explore the identified safety issues and welcomes further information that may be relevant, regardless of source. The HSIB will report any significant developments as the investigation progresses.