HSIB

HEALTHCARE SAFETY INVESTIGATION BRANCH

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Summary report

Undiagnosed cardiomyopathy in a young person with autism

Independent report by the
Healthcare Safety Investigation Branch I2018/026


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Providing feedback and comment on HSIB reports

At HSIB we welcome feedback on our investigation reports. The best way to share your views and comments is to email us at **enquiries@hsib.org.uk** We aim to provide a response to all correspondence within five working days.

www.hsib.org.uk/tell-us-what-you-think

About HSIB

The Healthcare Safety Investigation Branch (HSIB) conducts independent investigations of patient safety concerns in NHS-funded care across England. Most harm in healthcare results from problems within the systems and processes that determine how care is delivered. Our investigations identify the contributory factors that have led to harm or the potential for harm to patients.

The recommendations we make aim to improve healthcare systems and processes, to reduce risk and improve safety. Our organisation values independence, transparency, objectivity, expertise and learning for improvement. We work closely with patients, families and healthcare staff affected by patient safety incidents, and we never attribute blame or liability to individuals.

A note of acknowledgement

We are extremely grateful to the family whose experience is documented in this report. Their courage and openness has assisted the investigation greatly. The family expressed their wish that we use the name of their daughter, Alice, throughout relevant parts of this report.

Our investigations

Our team of investigators and analysts have diverse experience working in healthcare and other safety critical industries and are trained in human factors and safety science. We consult widely in England and internationally to ensure that our work is informed by appropriate clinical and other relevant expertise.

We undertake patient safety investigations through two programmes:

National investigations

Our national investigations can encompass any patient safety concern that occurred within NHS-funded care in England after 1 April 2017. We consider potential incidents or issues for investigation based on wide sources of information including that provided by healthcare organisations and our own research and analysis of NHS patient safety systems.

We decide what to investigate based on the scale of risk and harm, the impact on individuals involved and on public confidence in the healthcare system, and the learning potential to prevent future harm. We welcome information about patient safety concerns from the public, but we do not replace local investigations and cannot investigate on behalf of families, staff, organisations or regulators.

Our investigation reports identify opportunities for relevant organisations with power to make appropriate improvements though:

- 'Safety recommendations' made with the specific intention of preventing future, similar events; and
- 'Safety observations' with suggested actions for wider learning and improvement.

Our reports also identify 'safety actions' taken during an investigation to immediately improve patient safety.

We ask organisations subject to our recommendations to respond to us within 90 days. These responses are published on our website.

More information about our national investigations including in-depth explanations of our criteria, how we investigate, and how to refer a patient safety concern is available on our **website**.

Maternity investigations

From 1 April 2018, we have been responsible for all NHS patient safety investigations of maternity incidents which meet criteria for the **Each Baby Counts programme** (Royal College of Obstetricians and Gynaecologists, 2015) and also maternal deaths (excluding suicide). The purpose of this programme is to achieve learning and improvement in maternity services, and to identify common themes that offer opportunity for system-wide change. For these incidents HSIB's investigation replaces the local investigation, although the trust remains responsible for meeting the Duty of Candour and for referring the incident to us. We work closely with parents and families, healthcare staff and organisations during an investigation. Our reports are provided directly back to the families and to the trust. Our safety recommendations are based on the information derived from the investigations and other sources such as audit and safety studies, made with the intention of preventing future, similar events. These are for actions to be taken directly by the trust, local maternity network and national bodies.

Our reports also identify good practice and actions taken by the Trust to immediately improve patient safety.

Since 1 April 2019 we have been operating in all NHS Trusts in England.

We aim to make safety recommendations to local and national organisations for system-level improvements in maternity services. These are based on common themes arising from our trust-level investigations and where appropriate these themes will be put forward for investigation in the National Programme. More information about our maternity investigations is available on our **website**.

Executive Summary

Introduction

The objective of this investigation was to understand the context of magnetic resonance imaging (MRI) scanning under general anaesthetic and how care may be reasonably adjusted for patients with autism or learning disabilities. As an example, which we refer to as the 'reference event', we considered the experience of Alice, a teenage girl who had autism. Sadly, Alice died following her MRI scan under general anaesthetic.

The findings and conclusions of this investigation may be applicable to other non-invasive procedures carried out on patients who are under general anaesthetic.

The reference event

Alice, who was 14 years old, was being treated for growth hormone deficiency under the care of a consultant paediatrician, a community paediatrician and a regional specialist endocrinologist (an expert in conditions related to the glands and organs that produce hormones). Alice had a diagnosis of autism spectrum disorder which led to increased anxiety, and she also had learning difficulties.

Alice had begun having headaches and was referred for an MRI scan under general anaesthetic to rule out any serious illnesses. She attended a pre-anaesthetic assessment clinic a few weeks before attending for her scan.

On four occasions during the MRI scan, Alice required intervention to correct an abnormal heart rate. At the end of the scan she was found to be critically unwell. Alice was transferred to a specialist children's hospital, but sadly died a few days later.

At post-mortem, it was found that Alice had advanced hypertrophic cardiomyopathy (thickening of the heart walls) which had not previously been suspected, detected or diagnosed. Further tests conducted following Alice's post-mortem examination showed that she had a mitochondrial disorder which caused cardiac and skeletal muscle myopathy (muscle disease).

The national investigation

The reference event was referred to the Healthcare Safety Investigation Branch (HSIB) for potential investigation, and HSIB contacted the hospital where it had occurred. Following initial information gathering and evaluation against the HSIB patient safety risk criteria, HSIB's Chief Investigator authorised a national safety investigation.

The national investigation focused on: consent; pre-anaesthetic assessment services (assessments carried out by an appropriately trained clinician before a patient receives an anaesthetic); reasonable adjustments for autistic people and people with learning disabilities and learning difficulties; and preparing for unexpected adverse events relating to anaesthesia. This highlighted issues with the operationalisation of published practice guidance and opportunities to improve the experience of care and enhance safety. This has led to system-level safety recommendations being made to relevant bodies. The investigation identified:

- There is an opportunity to clarify the consent requirements for diagnostic imaging facilitated by a general anaesthetic.
- There is variation in the information given to patients regarding anaesthesia at the point of referral for an MRI scan under general anaesthetic.
- The observations and examinations to be routinely performed in pre-anaesthetic assessment are not defined nationally. The investigation found variation in the hospitals it visited.
- Children coming into hospital for an MRI scan who had been assessed as fit for anaesthetic were perceived as "well" by ward staff.
- Children with autism, learning disabilities and/or learning difficulties often find clinical environments distressing, which may be reflected in their physiological observations. This may result in diagnostic overshadowing, where problems such as autism (or a medical condition) are attributed as the cause of other new problems, rather than considering other underlying causes, thereby leaving other co-existing conditions potentially undiagnosed.
- Children with autism, learning disabilities or learning difficulties may benefit from reasonable adjustments being made when attending hospital.
- Electronic flagging systems can help staff identify patients who may benefit from reasonable adjustments. Hospital passports provide valuable information to assist with implementation of these adjustments.
- The model of care for learning disability nursing teams is not standardised nationally.

- There is an opportunity to enhance the existing published guidance available to assist clinicians involved in general anaesthetics to prepare for adverse events in the MRI scanning environment.
- Professional networks for anaesthetists provide the opportunity for shared learning and consensus regarding best practice.
- It is challenging to comply fully with the existing published standards for anaesthetic equipment used in MRI environments.

HSIB makes the following safety recommendations

Safety recommendation R/2020/079:
It is recommended that the Royal College of Anaesthetists convenes a working group to provide additional guidance regarding the responsibilities for obtaining consent for MRI and other non-invasive diagnostic and/or therapeutic procedures under general anaesthetic in children.

Safety recommendation R/2020/080:
It is recommended that the Royal College of Anaesthetists reviews standards for pre-assessment services, including their purpose, the required observations and examinations, and competencies of staff undertaking this work.

Safety recommendation R/2020/081:
It is recommended that NHS England and NHS Improvement strengthens its 'Learning disability improvement standards for NHS trusts' by including metrics which enable organisations to assess their progress against the outcomes for specialist learning disability teams.

Safety recommendation R/2020/082:
It is recommended that as part of the work to support the NHS Long Term Plan, NHS England and NHS Improvement should develop a role and competency framework for learning disability liaison nurses, to ensure that people with learning disabilities and autistic people receive optimal care which respects and protects their rights.

Safety recommendation R/2020/083:
It is recommended that NHSX develops a system for sharing care plans for patients with autism, learning disabilities or learning difficulties to enable reasonable adjustments to be made.

Safety recommendation R/2020/084:
It is recommended that NHSX develops a standardised care passport, which should include sections to support patients with autism, learning disabilities or learning difficulties.

Safety recommendation R/2020/085:
It is recommended that the Centre for Perioperative Care considers the remit of the National Safety Standards for Invasive Procedures (NatSSIPs) to cover the administration of general or regional anaesthesia for non-invasive diagnostic procedures.

Safety recommendation R/2020/086:
It is recommended that the Association of Anaesthetists reviews the dissemination and implementation of its 'Quick reference handbook' on managing adverse events during anaesthesia.

HSIB makes the following safety observation

Safety observation O/2020/065:
There are likely to be benefits for all organisations delivering anaesthesia to gain Anaesthesia Clinical Services Accreditation (ACSA) as this is likely to reduce unwarranted variation in practice.

HSIB notes the following safety actions

Safety action A/2020/030:
The recommendation for standardised anaesthetic equipment in the Royal College of Anaesthetists' 'Guidelines for the provision of anaesthetic services' is challenging within the MRI environment given the need for MR-safe/MR-conditional equipment. The Royal College of Anaesthetists has clarified this recommendation accordingly.

Safety action A/2020/031:
The Trust where the reference event took place has undertaken to resolve the errors with the clocks on the MRI scanner and anaesthetic monitoring equipment.

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Further information

More information about HSIB – including its team, investigations and history – is available at **www.hsib.org.uk**

If you would like to request an investigation then please read our **guidance** before submitting a safety awareness form.

@hsib_org is our Twitter handle. We use this feed to raise awareness of our work and to direct followers to our publications, news and events.

Contact us

If you would like a response to a query or concern please contact us via email using **enquiries@hsib.org.uk**

We monitor this inbox during normal office hours - Monday to Fridays (not bank holidays) from 0900hrs to 1700hrs. We aim to respond to enquiries within five working days.

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