Department of Health

From the Rt Hon Jeremy Hunt MP
Secretary of State for Health

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Mr A Tweddle
Senior Coroner
HM Coroner's Office
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14 APR 2014

Dear Mr. Tweddle,

Thank you for your letter following the inquest into the death of Nathan Douthwaite. In your report you conclude that the medical cause of death was abdominal compartment syndrome, perforated caecum and Hirschsprung's disease. I was very sorry to read of the events that led to the death of Mr Douthwaite and wish to extend my sincere sympathies to his family.

I understand Mr Douthwaite had suffered from severe constipation all his life and had frequent contact with health services. In 2000 when he was 6 years old he began treatment with a consultant paediatrician with a particular interest in paediatric constipation, who was involved in Mr Douthwaite's care until his death. In 2007 Mr Douthwaite was referred to a regional specialist paediatric surgeon, who considered Hirschsprung's disease exceptionally unlikely.

During 2010, Mr Douthwaite lost 17% of his body weight. His treating consultant made a diagnosis of functional constipation and did not diagnose Mr Douthwaite as suffering from Hirschsprung's disease. On 21 December 2010 Mr Douthwaite was admitted to hospital as an emergency and died the same day. An autopsy found a massive mega-colon, signs of abdominal compartment syndrome and perforation of the caecum. The cause of death is given as abdominal compartment syndrome, perforated caecum and Hirschsprung's disease.

The National Institute for Health and Care Excellence (NICE) published a clinical guideline covering Hirschsprung's disease in 2010. Rectal biopsy is given as a

method of diagnosis for Hirschsprung's disease. No rectal biopsy was ever performed on Mr Douthwaite as he did not meet the criteria for Hirschsprung's. Even under NICE's guidelines on constipation, published in May 2010, Mr Douthwaite's condition did not meet the criteria for rectal biopsy.

In evidence given to you by an independent consultant paediatric surgeon, it was suggested that there were more cases of undiagnosed Hirschsprung's disease in older children than is generally considered. It was also suggested that the relevant 2010 NICE guidelines should be reviewed for dealing with children with severe constipation.

You consider that had Mr Douthwaite undergone a rectal biopsy, it is likely that Hirschsprung's disease would have been diagnosed and he could then have been provided with appropriate treatment.

You therefore suggest:

- that NICE undertakes a review of the relevant guidelines;
- that County Durham and Darlington NHS Trust reviews its own practices and procedures in advance of a NICE review; and,
- that the Department of Health be aware of the circumstances of this case so that it can consider whether guidance should be issued pending the NICE review.

I am advised that this was a rare and unusual case as the majority of cases of Hirschsprung's Disease are diagnosed within the first few weeks of life by specialist consultants following referral from primary or secondary medical care.

I note that you have written to NICE about this case and that [redacted] has already responded. As you may be aware, NICE's clinical guidelines represent best practice and are based on the best available evidence and developed through wide consultation. NICE regularly reviews its published guidance to take account of the latest available evidence.

[redacted] has confirmed that NICE will shortly be starting a search for evidence to inform the review of its current clinical guidance on constipation in children and young people. He expects to publish the decision on whether the guidelines should be updated on NICE's website in June 2014. As mentioned above, NICE published its clinical guideline on the diagnosis and management of idiopathic childhood constipation in primary and secondary care in May 2010.

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The guidelines as they currently stand do not encourage rectal biopsy unless specific criteria are met and unless certain features are present. The clinical guideline recommends that a rectal biopsy is not performed unless any of the following clinical features of Hirschsprung's disease are or have been present:

- delayed passage of meconium (more than 48 hours after birth in term babies)
- constipation since first few weeks of life
- chronic abdominal distension plus vomiting
- family history of Hirschsprung's disease
- faltering growth in addition to any of the previous features.

Once NICE publishes guidance, health professionals and the organisations that employ them are expected to take the guidance fully into account when deciding how to diagnose and treat people. However, NICE guidance is not a substitute for the knowledge and skills of health professionals and treating clinicians must decide the most clinically appropriate course of action for each patient.

You ask the Department of Health to consider issuing interim guidance relating to this case pending the NICE review. NICE however has the statutory function of producing clinical guidelines. I consider that the expertise and responsibility lies with NICE in considering the need to update its existing guidance on constipation. Neither the Department of Health nor NHS England has the power or expertise to issue interim clinical guidelines as you propose.

NHS England has also considered whether a patient safety alert would be appropriate. However the circumstances of this case do not fall within the scope of an alert. The new patient safety alerting system requires that specific issues are identified for the purposes of creating an alert and it is important not to use alerts where guidelines are appropriate.

NHS England believes however that the circumstances of this tragic case merit dissemination to NHS learning networks to minimise the chances of any recurrence. To highlight the lessons to be learnt, NHS England will be placing an entry on its networks website.

I hope that this response is helpful and I am grateful to you for bringing the sad circumstances of Mr Douthwaite's death to my attention.

Yours sincerely

Jeremy Hunt

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