NHS Devon

NICE Update Bulletin: August 2023

This bulletin includes details of the following new and updated NICE guidance – click on the titles below for further details:

1. **Cipaglucosidase alfa with miglustat for treating late-onset Pompe disease TA912**
2. **Otitis media with effusion in under 12s NG233**
3. **Caesarean birth NG192 (UPDATE)**
4. **Venous thromboembolic diseases: diagnosis, management and thrombophilia testing NG158 (UPDATE)**
5. **Ectopic pregnancy and miscarriage: diagnosis and initial management NG126 (UPDATE)**
6. **Suspected cancer: recognition and referral NG12 (UPDATE)**
7. **Aortic valve reconstruction with glutaraldehyde-treated autologous pericardium for aortic valve disease IPG769**
8. **Quantitative faecal immunochemical testing to guide colorectal cancer pathway referral in primary care DG56**
9. **Genedrive MT-RNR1 ID Kit for detecting a genetic variant to guide antibiotic use and prevent hearing loss in babies: early value assessment HTE6 (UPDATE)**
10. **Current NICE Consultations**

Technology Appraisals (TAs)

Cipaglucosidase alfa with miglustat for treating late-onset Pompe disease TA912

Recommendations

Cipaglucosidase alfa (CIPA) plus miglustat is **recommended**, within its marketing authorisation, as an option for treating late-onset Pompe disease in adults. It is recommended only if the company provides it according to the commercial arrangement.

The Technology

Pompe disease, also known as glycogen storage disease type II or acid maltase deficiency, is a rare inherited genetic disorder caused by the mutation of the GAA gene which makes an enzyme called acid alpha-glucosidase, resulting in the deficiency of this enzyme. This leads to the progressive accumulation of glycogen, a sugar usually stored in multiple tissues including around the heart, skeletal muscles, respiratory muscles, vascular, gastrointestinal and nervous systems. The signs and symptoms of Pompe disease are directly related to the muscles affected. The respiratory, skeletal and cardiac muscles are most profoundly affected. Other symptoms include pain, mental fatigue and an impact on mental health.

Pompe disease is classified in two subtypes. The infantile onset and the late onset. The late onset usually presents from 1 year of age and is characterised by a progressive myopathy (with little or no cardiac involvement) which can lead to severe morbidity, respiratory failure and early mortality.

Current clinical management includes enzyme replacement therapy (ERT) with alglucosidase alfa which aims to replace the missing or malfunctioning enzyme. The decision to start treatment is usually based on a set of criteria including confirmed diagnosis and the patient should be symptomatic, have residual skeletal and respiratory muscle function and not have another advanced stage life-threatening condition. Supportive treatment is also needed and can include physiotherapist, occupational therapist, speech therapist and dietician.

Financial Factors

This technology is commissioned by NHS England.

NICE do not expect this guidance to have a significant impact on resources. This is because the technology is an alternative treatment option to existing enzyme replacement therapies. The overall cost of treatment for this patient group will be similar and the population size is small.

The company for cipaglucosidase alfa has a commercial arrangement. This makes it available to the NHS with a discount.

Highly Specialised Technology Guidance (HSTs)

None published this month.

NICE Guidelines (NGs)

Otitis media with effusion in under 12s NG233

This guidance updates and replaces CG60 (published February 2008)

This guideline covers identifying and managing otitis media with effusion (OME), also known as 'glue ear', in children younger than 12 years. It aims to improve hearing and quality of life in children with OME.

Caesarean birth NG192 (UPDATE)

This guideline covers when to offer and discuss caesarean birth, procedural aspects of the operation and care after caesarean birth. It aims to improve the consistency and quality of care for women and pregnant people who are thinking about having a caesarean birth or have had a caesarean birth in the past and are now pregnant again.

August 2023: NICE updated the recommendations on surgical opening technique for caesarean birth.

Venous thromboembolic diseases: diagnosis, management and thrombophilia testing NG158 (UPDATE)

This guideline covers diagnosing and managing venous thromboembolic diseases in adults. It aims to support rapid diagnosis and effective treatment for people who develop deep vein thrombosis (DVT) or pulmonary embolism (PE). It also covers testing for conditions that can make a DVT or PE more likely, such as thrombophilia (a blood clotting disorder) and cancer.

August 2023: NICE have updated recommendations on the use of Wells score and D-dimer in the diagnostic pathways for pulmonary embolism and deep vein thrombosis, following a review of the evidence for people with COVID-19. NICE also clarified the recommendation on the use of the pulmonary embolism rule-out criteria (PERC).

Ectopic pregnancy and miscarriage: diagnosis and initial management NG126 (UPDATE)

This guideline covers diagnosing and managing ectopic pregnancy and miscarriage in women with complications, such as pain and bleeding, in early pregnancy (that is, up to 13 completed weeks of pregnancy). It aims to improve how early pregnancy loss is diagnosed and the support women are given, to limit the psychological impact of their loss.

August 2023: NICE reviewed the evidence and made new and updated recommendations on medical management of miscarriage.

Suspected cancer: recognition and referral NG12 (UPDATE)

This guideline covers identifying children, young people and adults with symptoms that could be caused by cancer. It outlines appropriate investigations in primary care, and selection of people to refer for a specialist opinion. It aims to help people understand what to expect if they have symptoms that may suggest cancer.

August 2023: NICE updated the recommendations on criteria for faecal testing and referral for suspected colorectal cancer in line with DG56. The tables of symptoms and primary care investigation findings have been updated to reflect these changes.

NICE Rapid COVID-19 Guidelines

None published this month.

Public Health Guidelines

None published this month.

Antimicrobial Prescribing Guidelines

None published this month.

Social Care Guidelines

None published this month.

Interventional Procedures Guidance (IPGs)

Aortic valve reconstruction with glutaraldehyde-treated autologous pericardium for aortic valve disease IPG769

Recommendations

Aortic valve reconstruction with glutaraldehyde-treated autologous pericardium for aortic valve disease should be used **only in research**.

Further research could include the analysis of registry data or similar observational data to investigate patient selection and long-term outcomes, particularly the durability of the valve. Randomised controlled trials would also be useful to address uncertainties.

The Condition

Aortic valve disease is usually progressive, causing an increase in cardiac workload, left ventricular hypertrophy and heart failure. Symptoms can include palpitations, fatigue, shortness of breath, syncope and chest pain on exertion. Mortality rates are high in people who have symptoms.

Conventional treatment for a significantly diseased aortic valve is surgical replacement with an artificial prosthesis, or transcatheter aortic valve implantation (TAVI) with a biological prosthesis. Bioprosthetic and mechanical valves do not perform as well as valves made from the person's own tissue. Their durability is also limited (although mechanical valves last longer than bioprosthetic valves), which may be an issue for younger people. People with mechanical valves need lifelong anticoagulation. This increases the risk of haemorrhagic complications, particularly for older people and anyone with significant comorbidities or who wants to become pregnant. Aortic regurgitation can be treated by repairing the aortic valve with patches instead of replacing it.

Aortic valve reconstruction using glutaraldehyde-treated autologous pericardium is suitable for:

- people who cannot or do not want to take anticoagulation
- people with an aorta too narrow for a standard prosthetic valve
- young people who want to avoid long-term anticoagulation.

The Procedure

Under general anaesthesia, the heart is accessed using a full or partial sternotomy and the person is established on cardiopulmonary bypass. The heart is stopped with cardioplegic arrest. A section of the pericardium is removed and excess adipose tissue removed. The section of pericardium is treated with glutaraldehyde and rinsed with saline to avoid drying. The aorta is opened, the valve is inspected and the diseased valve cusps carefully removed. The intercommissural distances

are measured using Ozaki sizers and the treated pericardium is trimmed to the desired size and stitched to the aortic annulus to replace the removed valve leaflet(s). When aligned, the leaflets are stitched to the wall of the aorta to create a functional valve. The aorta is closed, the heart is de-aired and cardiopulmonary bypass is stopped. The circulation is restored and the chest is closed. The function of the valve is assessed intraoperatively by transoesophageal echocardiography.

Medical Technologies Guidance (MTGs)

None published this month.

Diagnostics Guidance (DGs)

Quantitative faecal immunochemical testing to guide colorectal cancer pathway referral in primary care DG56

This guidance replaces DG30 (published July 2017).

Recommendations

Quantitative faecal immunochemical testing (FIT) using HM-JACKarc or OC-Sensor is **recommended** to guide referral for suspected colorectal cancer in adults:

- with an abdominal mass, or
- with a change in bowel habit, or
- with iron-deficiency anaemia, or
- aged 40 and over with unexplained weight loss and abdominal pain, or
- aged under 50 with rectal bleeding and either of the following unexplained symptoms:
    - abdominal pain
    - weight loss, or
- aged 50 and over with any of the following unexplained symptoms:
    - rectal bleeding
    - abdominal pain
    - weight loss, or
- aged 60 and over with anaemia even in the absence of iron deficiency.

FIT should be offered even if the person has previously had a negative FIT result through the NHS bowel cancer screening programme. People with a rectal mass, an unexplained anal mass or unexplained anal ulceration do not need to be offered FIT before referral is considered.

Refer adults using a suspected cancer pathway referral (as outlined in NG12) for colorectal cancer if they have a FIT result of at least 10 micrograms of haemoglobin per gram of faeces.

For people who have not returned a faecal sample or who have a FIT result below 10 micrograms of haemoglobin per gram of faeces:

- safety netting processes should be in place
- referral to an appropriate secondary care pathway should not be delayed if there is strong clinical concern of cancer because of ongoing unexplained symptoms (for example, abdominal mass).

Clinicians should consider if people need additional help, information or support to return their sample.

Further research is recommended (see the section on further research) to:

- determine the clinical impact of using:
    - thresholds higher than 10 micrograms of haemoglobin per gram of faeces to guide referral
    - dual FIT
    - FIT in people aged under 40
- evaluate methods for improving access, uptake and return of FIT, especially in groups in which engagement is less likely
- determine how conditions or medicines that increase the risk of gastrointestinal bleeding affect the diagnostic accuracy of FIT.

Further research is recommended (see the section on further research) on the effectiveness of:

- FOB Gold
- IDK Hemoglobin ELISA
- IDK Hemoglobin/Haptoglobin Complex ELISA
- IDK TurbiFIT
- NS-Prime
- QuikRead go iFOBT.

The Tests

Colorectal cancer may be associated with a variety of symptoms, including blood in faeces. Small amounts of hidden blood in faeces can show that there is bleeding from the gastrointestinal tract, potentially from malignant growths on the inner lining of the large intestine. Several other conditions (such as inflammatory bowel disease) may also cause blood in faeces.

Faecal immunochemical testing (FIT) detects small amounts of blood in a faecal sample using antibodies specific to human haemoglobin. A positive FIT result alone cannot confirm a diagnosis of colorectal cancer. Further assessment using colonoscopy or CT colonography is needed to confirm diagnosis.

People referred to secondary care typically have further investigation using colonoscopy, or sometimes CT colonography. Clinicians have observed that many people on the suspected colorectal cancer referral pathway do not have any unusual findings at colonoscopy. So, using FIT could mean that people who are unlikely to have colorectal cancer may avoid colonoscopy and that people who are more likely to have colorectal cancer can be prioritised more effectively. Colonoscopy capacity is limited, and there are sometimes long waiting times. Using FIT could reduce the number of people referred for colonoscopy and so reduce the waiting times for people on non-urgent referral pathways.

The intervention in this guidance is quantitative FIT using specific thresholds of haemoglobin per gram of faeces to guide referral for people presenting to primary care with signs or symptoms suggestive of colorectal cancer.

The tests included in this guidance measure haemoglobin levels in faecal samples using either immunoturbidimetry or enzyme-linked immunosorbent assay (ELISA). Both methods use antibodies specific to human haemoglobin to bind to haemoglobin present in the faecal sample.

Financial Factors

This technology is commissioned by ICBs.

This guidance aligns with recommendations from the Association of Coloproctology of Great Britain & Ireland and the British Society of Gastroenterology and NHS England. The recommendations may lead to savings at a local level from a reduction in the number of colonoscopies performed. The level of these savings will be dependent on the extent to which NHS England's advice is currently being followed.

Health Technology Evaluations (HTE)

Genedrive MT-RNR1 ID Kit for detecting a genetic variant to guide antibiotic use and prevent hearing loss in babies: early value assessment HTE6 (UPDATE)

August 2023: NICE have developed tools and resources, in association with relevant stakeholders, to help organisations put this guidance into practice, including an evidence-generation plan. The evidence-generation plan discusses the prioritised evidence gaps and outcomes, ongoing studies, potential real-world data sources, and how remaining evidence gaps can be resolved through the design of real-world evidence studies.

NICE Quality Standards

None published this month.

Current NICE Consultations

Title/link	End date of consultation
Virtual Ward Platform Technologies for acute respiratory infections	01/09/2023
Dupilumab for treating prurigo nodularis [ID4054]	14/09/2023
Suspected cancer update	18/09/2023
Kidney Cancer	19/09/2023
Targeted temperature management to improve neurological outcomes after cardiac arrest	21/09/2023
Pharyngeal electrical stimulation for neurogenic dysphagia	21/09/2023
Diabetic retinopathy	27/09/2023