

## NHS England: Equality and Health Inequalities Impact Assessment (EHIA)

A completed copy of this form must be provided to the decision-makers in relation to your proposal. The decision-makers must consider the results of this assessment when they make their decision about your proposal.

**1. Name of the proposal (policy, proposition, programme, proposal or initiative)<sup>1</sup>:**

Clinical Commissioning Policy: Subcutaneous copper histidinate injections for presymptomatic neonates with Menkes disease (2105)

**2. Brief summary of the proposal in a few sentences**

Menkes disease is an inherited metabolic disorder of intestinal copper transport caused by mutations in the ATP7A gene on the X-chromosome, affecting male children. Menkes disease is a life-limiting, neurodegenerative disease mainly affecting boys who suffer drug-resistant seizures and a poor quality of life with frequent hospitalisations and significant healthcare resource utilisation during their life. Life expectancy is not normally greater than 3 years.

Copper histidinate is a formulation of copper that can be delivered underneath the skin by subcutaneous injection. Patients with Menkes disease have a defect in the copper transporter in the gut, which affects the normal absorption of copper from the diet. There is evidence that delivering copper histidinate by subcutaneous injection can reduce the symptoms caused by copper deficiency if given before symptoms have developed in the neonatal period.

**3. Main potential positive or adverse impact of the proposal for protected characteristic groups summarised**

Please briefly summarise the main potential impact (positive or negative) on people with the nine protected characteristics (as listed below). Please state **N/A** if your proposal will not impact adversely or positively on the protected characteristic groups listed below. Please note that these groups may also experience health inequalities.

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Protected characteristic groups	Summary explanation of the main potential positive or adverse impact of your proposal	Main recommendation from your proposal to reduce any key identified adverse impact or to increase the identified positive impact
<p><b>Age:</b> older people; middle years; early years; children and young people.</p>	<p>This policy is expected to have a positive impact on neonates (28 days and under) with Menkes disease as this policy suggests a treatment to stop symptoms and increase survival.</p> <p>This policy will have no impact on patients older than 28 days with Menkes disease as there is limited evidence of effectiveness for this treatment in this age group.</p>	<p>The policy reflects the best available evidence for treatment to be made available for those patients that would have positive outcomes. This means that children older than 28 days are not covered by this policy to be treated with copper histidinate.</p>
<p><b>Disability:</b> physical, sensory and learning impairment; mental health condition; long-term conditions.</p>	<p>This policy is expected to have a positive impact on neonates (28 days and under) with Menkes disease. Patients that are not treated commonly have brain and cognitive abnormalities with delayed growth and development, mainstream education is not normally feasible. Treatment with copper histidinate can reduce or stop these symptoms developing.</p>	<p>The policy is for a treatment option for presymptomatic neonates with Menkes disease. Provision of this treatment will provide an opportunity to slow down and stop the progression of the disease for patients to improve symptoms and their overall quality of life.</p>
<p><b>Gender Reassignment and/or people who identify as Transgender</b></p>	<p>There are no identified potential positive or adverse impacts of this policy on this group.</p>	<p>None</p>
<p><b>Marriage &amp; Civil Partnership:</b> people married or in a civil partnership.</p>	<p>There are no identified potential positive or adverse impacts of this policy on this group.</p>	<p>None</p>

Protected characteristic groups	Summary explanation of the main potential positive or adverse impact of your proposal	Main recommendation from your proposal to reduce any key identified adverse impact or to increase the identified positive impact
<b>Pregnancy and Maternity:</b> women before and after childbirth and who are breastfeeding.	This policy does not have a direct impact on this group. However, pregnant women who are identified as being at high-risk of a child with Menkes disease can be identified and genetic counselling can be offered, which can include treatment options for neonates including that proposed in this policy.	The policy is for the use of copper histidinate in presymptomatic neonates with Menkes disease. This requires diagnosis of the condition by up to 28 days following birth. Some of the eligible patients will be identified during pregnancy and genetic counselling can be offered to the families.
<b>Race and ethnicity</b> <sup>2</sup>	There are no identified potential positive or adverse impacts of this policy on this group.	None
<b>Religion and belief:</b> people with different religions/faiths or beliefs, or none.	There are no identified potential positive or adverse impacts of this policy on this group.	None
<b>Sex:</b> men; women	Menkes disease is an X-linked recessive disorder, so primarily affects males. This policy is expected to have a positive impact on male neonates (28 days and under) with Menkes disease as this policy suggests a treatment to stop symptoms and increase survival.	Due to the X-linked inheritance of Menkes disease, affected children are more likely to be male and therefore it is more likely that males will be treated under this policy. As the majority of females with ATP7A mutations are clinically unaffected, it is not appropriate to administer this treatment to females routinely. This does potentially mean that the exceptionally rare affected female patient would not qualify for treatment, as by the time their clinical

<sup>2</sup> Addressing racial inequalities is about identifying any ethnic group that experiences inequalities. Race and ethnicity includes people from any ethnic group incl. BME communities, non-English speakers, Gypsies, Roma and Travelers, migrants etc. who experience inequalities so includes addressing the needs of BME communities but is not limited to addressing their needs, it is equally important to recognise the needs of White groups that experience inequalities. The Equality Act 2010 also prohibits discrimination on the basis of nationality and ethnic or national origins, issues related to national origin and nationality.

Protected characteristic groups	Summary explanation of the main potential positive or adverse impact of your proposal	Main recommendation from your proposal to reduce any key identified adverse impact or to increase the identified positive impact
		status is realised it would be outside of 28 days and outside of the scope of this policy to treat. If a family who has had a previously affected female patient wishes to test a future female baby biochemically to see if they are likely to be affected, this may be possible under this policy.
<b>Sexual orientation:</b> Lesbian; Gay; Bisexual; Heterosexual.	There are no identified potential positive or adverse impacts of this policy on this group.	None

#### 4. Main potential positive or adverse impact for people who experience health inequalities summarised

Please briefly summarise the main potential impact (positive or negative) on people at particular risk of health inequalities (as listed below). Please state **N/A** if your proposal will not impact on patients who experience health inequalities.

Groups who face health inequalities <sup>3</sup>	Summary explanation of the main potential positive or adverse impact of your proposal	Main recommendation from your proposal to reduce any key identified adverse impact or to increase the identified positive impact
<b>Looked after children and young people</b>	There are no identified potential positive or adverse impacts of this policy on this group.	None
<b>Carers of patients:</b> unpaid, family members.	This policy is expected to have a positive impact on carers of patients with Menkes disease. Patients that are not treated commonly have brain and cognitive	The policy reflects the best available evidence for treatment to be made available for those patients that would have positive outcomes. The policy recommends referring patients and carers to patient

<sup>3</sup> Please note many groups who share protected characteristics have also been identified as facing health inequalities.

<b>Groups who face health inequalities<sup>3</sup></b>	<b>Summary explanation of the main potential positive or adverse impact of your proposal</b>	<b>Main recommendation from your proposal to reduce any key identified adverse impact or to increase the identified positive impact</b>
	abnormalities with delayed growth and development, mainstream education is not normally feasible. Treatment with copper histidinate can reduce or stop these symptoms developing. It is noted that administration of the treatment can be difficult for carers psychologically as the injections can be painful.	support organisations, which may mitigate the psychological stress of administering painful injections. Further input from psychology services should be considered by the multidisciplinary team for carers of patients that are struggling. Carers will also be supported prior to discharge with training on how to do this and will not be discharged until confident and competent with the injections.
<b>Homeless people.</b> People on the street; staying temporarily with friends /family; in hostels or B&Bs.	There are no identified potential positive or adverse impacts of this policy on this group.	None
<b>People involved in the criminal justice system:</b> offenders in prison/on probation, ex-offenders.	There are no identified potential positive or adverse impacts of this policy on this group.	None
<b>People with addictions and/or substance misuse issues</b>	There are no identified potential positive or adverse impacts of this policy on this group.	None
<b>People or families on a low income</b>	This policy suggests follow-up with specialist multidisciplinary teams (MDTs). There are approximately fifteen specialist centres and within that there are some larger centres, to which patients with ultra-rare conditions may be referred. Therefore, treatment may be initiated in these centres with follow-up requiring travel to the closest of these centres.	Follow-up may be able to take place virtually where appropriate, with blood tests and imaging being undertaken at local healthcare centres (at GP practices or local hospitals). Due to the rarity of the condition, it is not possible to increase the number of centres.
<b>People with poor literacy or health Literacy:</b> (e.g. poor understanding	Carers will be required to give twice daily injections to the patient for the first year of life. These carers must be adequately	Carers with poor literacy or health literacy may receive further help from staff on the wards and the patient may not be discharged until the carers are

Groups who face health inequalities <sup>3</sup>	Summary explanation of the main potential positive or adverse impact of your proposal	Main recommendation from your proposal to reduce any key identified adverse impact or to increase the identified positive impact
of health services poor language skills).	trained on home administration of the injections, which may be more difficult for people with poor literacy or health literacy.	adequately trained in the administration of the injections. Further support could be offered including pictorial aids and verbal instructions as required on a person-by-person basis. As the dose of copper histidinate is fixed these aids mean it can be taught to those with poor literacy skills, although it is recognised that these carers will need significantly more support prior to discharge.
<b>People living in deprived areas</b>	This policy suggests follow-up with specialist multidisciplinary teams (MDTs). There are approximately fifteen specialist centres and within that there are some larger centres, to which patients with ultra-rare conditions may be referred. Therefore, treatment may be initiated in these centres with follow-up requiring travel to the closest of these centres; this will be at least every 6 months.	Follow-up may be able to take place virtually where appropriate, with blood tests and imaging being undertaken at local healthcare centres (at GP practices or local hospitals). Due to the rarity of the condition, it is not possible to increase the number of centres.
<b>People living in remote, rural and island locations</b>	This policy suggests follow-up with specialist multidisciplinary teams (MDTs). There are approximately fifteen specialist centres and within that there are some larger centres, to which patients with ultra-rare conditions may be referred. Therefore, treatment may be initiated in these centres with follow-up requiring travel to the closest of these centres; this will be at least every 6 months.	Follow-up may be able to take place virtually where appropriate, with blood tests and imaging being undertaken at local healthcare centres (at GP practices or local hospitals). Due to the rarity of the condition, it is not possible to increase the number of centres.

<b>Groups who face health inequalities<sup>3</sup></b>	<b>Summary explanation of the main potential positive or adverse impact of your proposal</b>	<b>Main recommendation from your proposal to reduce any key identified adverse impact or to increase the identified positive impact</b>
<b>Refugees, asylum seekers or those experiencing modern slavery</b>	There are no identified potential positive or adverse impacts of this policy on this group.	None
<b>Other groups experiencing health inequalities (please describe)</b>	There are no further identified potential positive or adverse impacts of this policy on any other groups experiencing health inequalities.	None

## 5. Engagement and consultation

a. Have any key engagement or consultative activities been undertaken that considered how to address equalities issues or reduce health inequalities? Please place an x in the appropriate box below.

<b>Yes X</b>	<b>No</b>	<b>Do Not Know</b>
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b. If yes, please briefly list up the top 3 most important engagement or consultation activities undertaken, the main findings and when the engagement and consultative activities were undertaken.

<b>Name of engagement and consultative activities undertaken</b>	<b>Summary note of the engagement or consultative activity undertaken</b>	<b>Month/Year</b>
<b>1</b> Policy working group	The policy working group that has developed the policy is made up of specialist clinicians, a patient public voice representative, a public health consultant, a pharmacist and a commissioner to offer a wide range of opinions and background	Throughout the policy process
<b>2</b> Stakeholder testing	A two week period of stakeholder testing was undertaken	08.03.2022 and 23.03.2022

**6. What key sources of evidence have informed your impact assessment and are there key gaps in the evidence?**

<b>Evidence Type</b>	<b>Key sources of available evidence</b>	<b>Key gaps in evidence</b>
<b>Published evidence</b>	An external review of available clinical evidence was undertaken to inform this policy.	There was no evidence on cost effectiveness or on any subgroups of patients who may benefit more than others from early treatment delivered prior to symptoms developing. There was also no evidence available for health related quality of life, number of hospital attendances/admissions, requirement for anti-convulsant medication and development of bladder diverticulae.
<b>Consultation and involvement findings</b>	There were no responses to stakeholder testing, not usual when the patient and key clinical groups are members of the PWG.	
<b>Research</b>	No pending research is known	N/A
<b>Participant or expert knowledge</b> For example, expertise within the team or expertise drawn on external to your team	Policy working group is formulated of national clinical experts in the field, along with a public health consultant and a patient public voice representative from a patient organisation.	

**7. Is your assessment that your proposal will support compliance with the Public Sector Equality Duty? Please add an x to the relevant box below.**

	Tackling discrimination	Advancing equality of opportunity	Fostering good relations
The proposal will support?			
The proposal may support?			
Uncertain whether the proposal will support?	X	X	X



**8. Is your assessment that your proposal will support reducing health inequalities faced by patients?** Please add an x to the relevant box below.

	Reducing inequalities in access to health care	Reducing inequalities in health outcomes
The proposal will support?		
The proposal may support?		
Uncertain if the proposal will support?	X	X

**9. Outstanding key issues/questions that may require further consultation, research or additional evidence.** Please list your top 3 in order of priority or state N/A

Key issue or question to be answered		Type of consultation, research or other evidence that would address the issue and/or answer the question
1	N/A	
2		
3		

**10. Summary assessment of this EHIA findings**

This policy suggests the use of copper histidinate injections for presymptomatic neonates with Menkes disease where no treatment currently exists. This is expected to have a positive impact on those patients which are eligible. There are some potential adverse impacts of the policy in that treatment must be initiated before 28 days of age. Patients identified as having Menkes disease after 28 days following birth would not be eligible for treatment under the policy, as there is limited evidence of efficacy of copper histidinate in this group. Furthermore, follow-up is required at one of around 15 specialist centres and within that there are some larger centres, to which patients with ultra-rare conditions may be referred. Therefore, treatment may be initiated in these centres with follow-up requiring travel to the closest of these centres, which may be difficult for patients in remote, rural or island locations or low-income

families. Due to the rarity of the condition, it is not possible to increase the number of centres. Virtual follow-up may be available in certain circumstances.

**11. Contact details re this EHIA**

Team/Unit name:	Women and Children National Programme of Care
Division name:	Specialised Commissioning, NHS England
Directorate name:	Chief finance officer
Date EHIA agreed:	
Date EHIA published if appropriate:	