Positional Magnetic Resonance Imaging for People With Ehlers-Danlos Syndrome or Suspected Craniovertebral or Cervical Spine Abnormalities: OHTAC Recommendation

HEALTH QUALITY ONTARIO

ONTARIO HEALTH TECHNOLOGY ADVISORY COMMITTEE RECOMMENDATIONS
Given the current absence of evidence for the utility of positional magnetic resonance imaging (pMRI), OHTAC recommends that pMRI not be funded or considered generally accepted practice for the diagnosis or management of (a) spinal or craniovertebral abnormalities among individuals with Ehlers-Danlos Syndrome, or (b) major craniovertebral or cervical spine abnormalities among symptomatic individuals.

BACKGROUND
Ehlers-Danlos syndrome and other congenital, developmental, or acquired disorders can lead to abnormalities within the spine and craniovertebral junction (CVJ). Appropriate imaging and diagnosis is needed to determine patient management and need for complex surgery. Positional MRI allows imaging of the spine or CVJ with patients in upright, weight-bearing positions as well as in combination with dynamic maneuvers. Imaging with pMRI could allow diagnosticians to better detect spinal or CVJ abnormalities than recumbent MRI or a combination of other available imaging modalities might allow.

REVIEW OF THE EVIDENCE
Research Questions
Health Quality Ontario conducted an evidence-based analysis (1) to answer the following research questions:

- **Question 1**—What is the diagnostic impact and clinical utility of pMRI in the assessment of craniovertebral or spinal abnormalities among people with EDS relative to currently available diagnostic modalities?
- **Question 2**—What is the diagnostic impact and clinical utility of pMRI in the assessment of major craniovertebral or cervical spinal abnormalities among symptomatic people relative to currently available diagnostic modalities?

For a fuller discussion and the evidence related to these recommendations, please see the related report. (1)

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(1) Specific major cervical or craniovertebral spinal abnormalities or consequences were defined on the basis of expert consultation: they included craniovertebral or cervical spine instability (atlanto-axial instability; vertical subluxation, basilar invagination, or cranial settling; and subaxial instability); cervical spine stenosis; Chiari malformation; and cervical spine compression or brainstem compression.

Main Findings
- We did not identify any evidence that assessed the diagnostic impact or clinical utility of pMRI in the assessment of craniovertebral or spinal abnormalities among people with EDS relative to currently available diagnostic modalities.
- We did not identify any evidence that assessed the diagnostic impact or clinical utility of pMRI in the assessment of major craniovertebral or cervical spine abnormalities among symptomatic people relative to currently available diagnostic modalities.

OHTAC DELIBERATIONS
Health Quality Ontario has developed a decision-making framework to help guide deliberation and support the development of OHTAC recommendations regarding the uptake, diffusion, distribution, or removal of health interventions in Ontario. Table 1 provides a summary of the decision determinants for this recommendation.
### Table A1: Decision Determinants for Positional Magnetic Resonance Imaging

<table>
<thead>
<tr>
<th>Decision Criteria</th>
<th>Subcriteria</th>
<th>Decision Determinants Considerations</th>
</tr>
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<tbody>
<tr>
<td>Overall clinical benefit</td>
<td>Effectiveness</td>
<td>• No evidence was identified that assessed the diagnostic impact or clinical utility of pMRI in the diagnosis or management of craniovertebral or spinal abnormalities among people with EDS.</td>
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<tr>
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<td>Safety</td>
<td>MRI is contraindicated for patients with implanted ferromagnetic devices. Additionally, imaging patients in the upright position with dynamic maneuvers could further aggravate patient pain and symptoms.</td>
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<tr>
<td></td>
<td>Burden of Illness</td>
<td>• Overall prevalence of EDS is estimated at 1 in 5,000. The proportion of people with EDS experiencing abnormalities of the spine or CVJ is unknown, with minimal evidence published to date.</td>
</tr>
<tr>
<td></td>
<td>Need</td>
<td>• Prevalence of nontraumatic upper cervical spine instability and associated abnormalities in the general symptomatic population is unknown, and differs among various patient populations that are predisposed to these abnormalities.</td>
</tr>
</tbody>
</table>
| Consistency with expected societal and ethical values | Societal values | No formal review of expected societal and ethical values was conducted. Symptoms of CVJ abnormalities manifest in a multitude of ways, the causes of which are often difficult to diagnose and subsequently treat. Despite a lack of evidence for or against effectiveness and safety, imaging with pMRI could reassure patients and add confidence in their assessment.
|                                        | Ethical values                                                             | Major cervical spine or CVJ abnormalities can result in severe pain, morbidity, and potentially irreversible neurologic compromise. Appropriate diagnosis and management of these abnormalities is needed in order to determine appropriate treatment including need for complex spinal surgery. |
| Value for money                         | Economic evaluation                                                        | Unclear. Insufficient evidence to addresses this question.                                                                                                                                                                             |
| Feasibility of adoption into health system | Economic feasibility                                                      | Unclear                                                                                                                                                                                                                          |
|                                        | Organizational feasibility                                                 | There are currently no pMRI machines in Ontario. Acquiring a machine would require installation specifications similar to those of conventional MRI devices. Hospital staff would need to be trained on the use of corresponding software, scanning protocols, and patient positioning. |

**Abbreviations:** CVJ, craniovertebral junction; EDS, Ehlers-Danlos syndrome; MRI, magnetic resonance imaging; pMRI, positional magnetic resonance imaging.

*Anticipated or assumed common ethical and societal values held in regard to the target condition, target population, and/or treatment options. Unless there is evidence from scientific sources to corroborate the true nature of the ethical and societal values, the expected values are considered.
REFERENCES

DISCLAIMER
The analysis may not have captured every relevant publication and relevant scientific findings may have been reported since the development of this recommendation. This report may be superseded by an updated publication on the same topic.

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