Quantification of Optic Canal Stenosis Reversal after Hematopoietic Stem Cell Transplantation in Pediatric Patients with Osteopetrosis as Measured on CT and MR Imaging.

Kunal M. Patel¹, David R. Nascene¹,²

¹ Department of Radiology, University of Minnesota Medical Center, Minneapolis, Minnesota; ²Department of Radiology, Amplatz Children’s Hospital, Minneapolis, Minnesota.

REFERENCES:


CONCLUSION:

1. Reversal of optic canal stenosis occurs after HSCT; however, values do not equal age-matched controls.
2. Measurement of optic canal diameters prior to HSCT on neuroimaging studies of Osteopetrosis patients serve as a baseline for comparison to post HSCT values.
3. Long-term study of optic canal diameters as patients age may be beneficial to evaluate whether values eventually match age-matched controls.
4. Clinical correlative data regarding restoration of visual function would also be of value and could be performed in this long-term study.

PURPOSE:

1. Osteopetrosis: Rare devastating inherited disorder due to osteoclastic dysfunction.
2. Common symptom: Optic canal stenosis → visual loss.
3. Hematopoietic Stem Cell Transplantation (HSCT): only known durable treatment of Osteopetrosis¹.
4. Visual loss is noted to improve after HSCT but the mechanism remains unclear.
5. Most common purported mechanism: Reversal of optic canal stenosis with CN II decompression.
6. Efficacy of HSCT in reversing optic canal stenosis is not well studied with only one case report available².

MATERIALS AND METHODS:

- At our institution, 24 patients with severe Osteopetrosis were treated with HSCT in the past 33 years.
- 7 patients had suitable cross-sectional neuroimaging studies:
  - Before HSCT (average: 73 ± 68 days) – AND –
  - After HSCT (average: 185 ± 88 days)
- 2 adult patients with delayed disease onset were not included.
- We measured the optic canal diameter on all CT and MRI studies available on:
  - 5 Osteopetrosis patients (mean age @ BMT: 0.75 ± 0.24 yrs)
  - 17 age matched control patients (mean age: 1.04 ± 0.40 yrs)
- Comparisons of pre- and post- HSCT values were performed using a student’s t-test.

RESULTS:

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<tr>
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<th>O.C. diameter (mm)</th>
<th>P-value</th>
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<tbody>
<tr>
<td><strong>OSTEOPETROSIS</strong></td>
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<tr>
<td>Pre-HSCT:</td>
<td>1.93 ± 0.50</td>
<td>0.01</td>
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<tr>
<td>Post-HSCT:</td>
<td>2.53 ± 0.54</td>
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<tr>
<td><strong>CONTROLS</strong></td>
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<td>3.86 ± 0.49</td>
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Table 1: There was a 31% increase in Optic Canal (O.C.) diameters of Osteopetrosis patients after HSCT. Values did not match age-matched controls.

Figure 1: Sample comparison of optic canal diameter on pre HSCT (TOP) and post HSCT (BOTTOM) on axial CT images in the same patient.

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CORRESPONDENCE:
KUNAL PATEL – patel085@umn.edu.