A 17-year-old boy was diagnosed with congenital lipomatous overgrowth with vascular, epidermal, skeletal, and spinal anomalies (CLOVES) syndrome, mainly affecting his right face, brain, and trunk (MIM#612918) (figure, A–C). Brain MRI revealed right hemimegalencephaly with extensive temporo-parieto-occipital cortical dysplasia (figure 1, D1-4). He developed neonatal drug-resistant seizures requiring right hemispherectomy at 15 months. He has left hemiparesis and intellectual disability. CLOVES syndrome is a segmental overgrowth syndrome associated with somatic hyperactivating mutations in PIK3CA, belonging to the mammalian target of rapamycin signaling pathway. Genetic testing on buccal swab revealed a pathogenic somatic missense mutation in PIK3CA (NM_006218.4:c.1624G>A, p.Glu542Lys) at an alternate allele frequency of 4.5%, which was absent in blood.

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**Disclosure**
The authors report no disclosures relevant to the manuscript. Go to Neurology.org/N for full disclosures.

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### Appendix

#### Authors

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<tr>
<th>Name</th>
<th>Location</th>
<th>Contribution</th>
</tr>
</thead>
<tbody>
<tr>
<td>Meagan Collins, BS</td>
<td>Child Health and Human Development Program, Research Institute of the McGill University Health Centre, Montreal, Quebec, Canada CHU Sainte Justine Research Center, Université de Montréal, Quebec, Canada</td>
<td>Drafting/revising the manuscript, study concept or design, accepts responsibility for conduct of research and final approval, acquisition of data, and study supervision</td>
</tr>
<tr>
<td>Eric Krochmalnek, BS</td>
<td>Child Health and Human Development Program, Research Institute of the McGill University Health Centre, Montreal, Quebec, Canada</td>
<td>Drafting/revising the manuscript, study concept or design, accepts responsibility for conduct of research and final approval, acquisition of data, and study supervision</td>
</tr>
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<tr>
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<th>Location</th>
<th>Contribution</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sarah Alsubhi, MD</td>
<td>Division of Pediatric Neurology, Department of Pediatrics, McGill University, Montreal, QC, Canada</td>
<td>Drafting/revising the manuscript, study concept or design, accepts responsibility for conduct of research and final approval, acquisition of data, and study supervision</td>
</tr>
<tr>
<td>Myriam Srour, MD, PhD</td>
<td>Child Health and Human Development Program, Research Institute of the McGill University Health Centre, Montreal, Quebec, Canada Division of Pediatric Neurology, Department of Pediatrics, McGill University, Montreal, Quebec, Canada</td>
<td>Drafting/revising the manuscript, study concept or design, accepts responsibility for conduct of research and final approval, acquisition of data, and study supervision</td>
</tr>
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</table>

### References

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