Hydrocephalus in an achondroplastic child treated by venous decompression at the jugular foramen

Case report

TRYGGVE LUNDAR, M.D., SØREN JACOB BAKKE, M.D., AND HELGE NORNES, M.D.

Departments of Neurosurgery and Neuroradiology, The National Hospital, University of Oslo, Oslo, Norway

A 10-month-old child with achondroplasia with progressive head enlargement, ventriculomegaly, and wide subarachnoid spaces over the hemispheres was referred for evaluation. A steady-state lumbar infusion test revealed increased cerebrospinal fluid (CSF) outflow resistance (14 mm Hg/ml/min), and intra-arterial digital subtraction angiography (DSA) demonstrated bilateral venous outflow obstruction due to stenosis of the jugular foramen. Surgical decompression by opening the right jugular foramen relieved the clinical signs of intracranial hypertension. During the following year, the patient's head enlargement was moderate with relative normalization of size. Repeat DSA demonstrated improved venous runoff on the right side, and a steady-state lumbar infusion test demonstrated reduced CSF outflow resistance (10 mm Hg/ml/min). Venous decompression is causal therapy and may prove to be preferable to shunting in children with hydrocephalus and bilateral stenosis of the jugular foramen.

KEY WORDS • achondroplasia • hydrocephalus • venous decompression • jugular foramen

ENLARGED head and ventriculomegaly is a common finding in children with achondroplasia. In a recent study, Steinbok, et al.,2 demonstrated evidence of active hydrocephalus as well as bilateral stenosis of the jugular foramen in five achondroplastic children. This report describes relief of clinical symptoms of increased intracranial pressure and improved cerebrospinal fluid (CSF) absorptive capacity after decompression of the vein in the right jugular foramen of a child with bilateral stenosis.

Case Report

This achondroplastic boy was born 5 weeks prematurely and was referred to our neurosurgical department at the age of 9 months for evaluation of progressive head enlargement (head circumference 48 cm, 35 cm at birth). He presented with a widened and tense anterior fontanel, markedly distended scalp veins, and a pathological percussion sound. Repeated computerized tomography (CT) scans revealed progressive ventriculomegaly and widened subarachnoid spaces over both hemispheres (Fig. 1 left and center). A steady-state lumbar infusion test under general anesthesia (fentanyl/N₂O) with moderate hyperventilation (PaCO₂ 4 kPa) disclosed an opening pressure (Po) of 10 mm Hg and a plateau pressure (Pp) of 31 mm Hg during a 1.5-ml/min infusion. According to the formula,2 Ro = (Pp – Po)/infusion rate, the outflow resistance (Ro) was estimated as 14 mm Hg/ml/min, which is pathologically increased. Intra-arterial digital subtraction angiography (DSA) demonstrated severe bilateral stenosis (75% to 90% luminal reduction) of the vein at the level of the jugular foramen (Fig. 2 left).

Operation. One month later, the head circumference had increased to 48.8 cm (Fig. 3) and a right-sided decompression procedure was performed through a paramedian suboccipital approach with the aid of magnifying glasses. After identification of the junction between the transverse and sigmoid sinuses, the latter was followed anteriorly to the jugular foramen, which was widely opened posteriorly. The kinking of the vein by a ridge of bone was even more impressive than had appeared on the angiogram.

Postoperative Course. Fontanel tension as well as scalp vein distention were reduced immediately after the operation and the child's general condition im-
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Fig. 1. Computerized tomography scans demonstrating ventriculomegaly and enlarged subarachnoid spaces at 6 months of age (left), preoperatively at 9 months (center), and at 14 months (4 months postoperatively) with slightly reduced ventricular size (right).

proved markedly. During the subsequent weeks his head circumference was reduced by 1 cm and thereafter increased more slowly with relative normalization following the 75th percentile (Fig. 3). Four months after the operation, repeat DSA demonstrated reduced venous stenosis at the right jugular foramen with improved venous runoff (Fig. 2 right). A repeat steady-state infusion test disclosed reduced CSF outflow resistance (10 mm Hg/ml/min). The follow-up CT scan showed a slight reduction in ventricular size (Fig. 1 right).

Discussion

Impaired cerebral venous outflow due to bilateral stenosis of the jugular foramen has been demonstrated in individuals with achondroplasia. Steinbok, et al., described hydrocephalus as well as significant pressure gradients across the stenosed jugular foramina in the four children they studied. In our patient, progressive ventriculomegaly was accompanied by marked clinical signs of hydrocephalus and pathologically increased CSF outflow resistance. The implantation of shunt systems in individuals with achondroplasia has been

Fig. 2. Left: Intra-arterial digital subtraction angiography (DSA) demonstrating impaired venous runoff due to bilateral high-grade venous stenosis (75% to 90% luminal obstruction) at the level of the jugular foramen. Right: Postoperative DSA showing improved venous runoff on the decompressed right side.
compression of a venous outflow obstruction may reduce the hydrocephalic problem more gradually than ventricular fluid diversion to the peritoneal cavity in such patients. The surgical decompression described here and its effect on CSF circulation is probably more like the slow reduction in ventricular size seen after internal shunting procedures (third ventriculostomy or ventriculocisternostomy).

Further follow-up studies and more experience are needed, however, before this can be considered an established treatment modality in selected cases. In this particular patient, the clinical condition appeared satisfactory 15 months after surgery. If clinical signs of progressive hydrocephalus should reappear, repeat or contralateral bone decompression around the vein might be considered.

References


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Address reprint requests to: Trygve Lundar, M.D., Department of Neurosurgery, Rikshospitalet, N-0027, Oslo 1, Norway.