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Usefulness of MR Imaging in Children without Characteristic Clinical Findings of Duane's Retraction Syndrome

Jae Hyoung Kim and Jeong-Min Hwang

BACKGROUND AND PURPOSE: Duane's retraction syndrome (DRS) consists of a congenital abduction deficit of the eyeball accompanied by retraction of the globe on attempted adduction and by upshoots or downshoots of the affected eye on adduction. These characteristic diagnostic signs of DRS, however, might not be manifested in early childhood. We evaluated the usefulness of MR imaging as a diagnostic tool in such cases.

METHODS: Thin-section gradient-echo imaging at the brain stem level was performed in two pediatric patients with only abduction deficit and 10 control children. Imaging findings were analyzed focused on the presence or absence of the abducens nerve.

RESULTS: The abducens nerve on the affected side was absent in three of three affected eyes in two patients. The right and left abducens nerves were well identified in all 10 control subjects.

CONCLUSION: MR imaging is useful for the differential diagnosis of abduction deficit of the eyeball in pediatric patients. The absence of the abducens nerve suggests DRS strongly in children with abduction deficit.

Duane's retraction syndrome (DRS) consists of a congenital abduction deficit accompanied by retraction of the globe on attempted adduction and by upshoots or downshoots of the affected eye on adduction (1). A few pathologic examinations revealed that the abducens nucleus and nerve corresponding to the side with the abduction deficit is absent or hypoplastic from the brain stem in the patients with DRS (2–4). The incidence of severe retraction on adduction and the presence of upshoots and downshoots, which are clinically considered diagnostic signs of DRS in patients with abduction deficit, however, were reported to be significantly lower in children with DRS than in adults (5, 6). Thus, many children with DRS present only with an abduction deficit before later developing these typical diagnostic signs of DRS, such as globe retraction or upshoots or downshoots. Common differential diagnoses in children with only abduction deficit include DRS, 6th nerve palsy, and infantile esotropia. Two recent articles reported the absence of the abducens nerve on MR imaging in some patients with DRS (7, 8).

We recently noted the absence of the abducens nerve in two children with only congenital abduction deficit and without other characteristic diagnostic signs of DRS. We hypothesized that thin-section MR imaging would depict the abducens nerve and thereby help differentiate DRS from other diagnoses. In this study, we also performed thin-section MR imaging in 10 unaffected children and evaluated the presence or absence of the abducens nerve.

Methods

Two children with only abduction deficit among the characteristic findings of DRS between May 2003 and April 2004 were included in this study. The first patient, a 6-year-old boy, was referred for congenital abduction deficit of the right eye. He had orthotropia in the primary position and at left gaze. At right gaze, severe esotropia of 45 prism diopters (Δ) was found with a marked abduction deficit of -4 out of a grading from -4 (maximal limitation) to 0 (full rotation) in the right eye. The second patient, a 4-year-old girl, was referred for esotropia. She had mild intermittent esotropia of 12 Δ in the primary position and a marked abduction deficits of -4 in both eyes. Neither patient showed any definite retraction of the globe on attempted adduction or upshoots or downshoots of the right eye on adduction. Medical histories of both patients were noncontributory.

Ten children (age range, 4–10 years; mean age, 7 years) were included as control subjects to evaluate the accuracy of thin-section MR imaging to depict the abducens nerve. They were selected from the patients who underwent brain MR imaging for headache without other neurologic symptoms, including ocular movement abnormality. Institutional review

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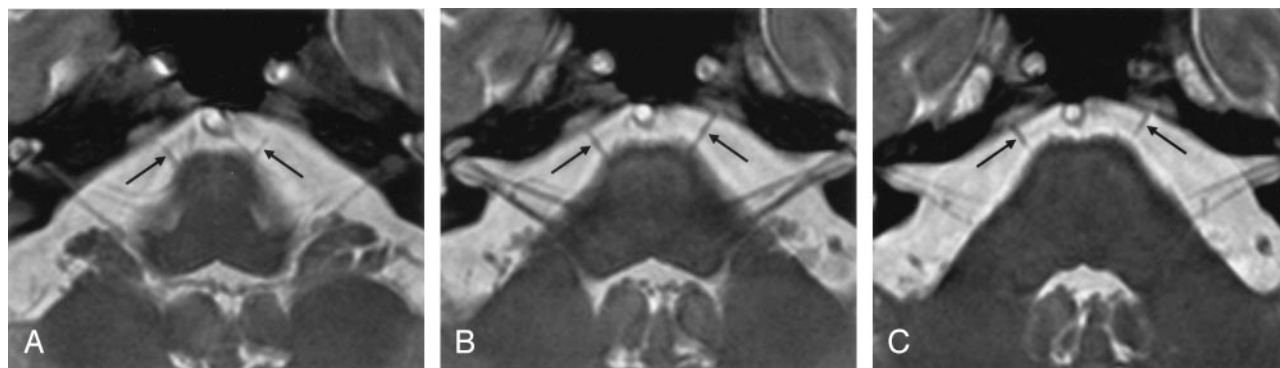


FIG 1. Normal abducens nerves.

A–C, Three axial MR images show the entire path of the right and left abducens nerves (arrows) as linear dark structures emerging from the pontomedullary sulcus (A), coursing in the superior oblique direction (B), and finally entering into the clivus (C).

board approval was not required for this study. Informed consent was obtained from parents of all patients and control subjects.

MR imaging was conducted by using a 1.5T system (Gyrosan Intera; Philips, Best, the Netherlands) with a SENSE head coil and a 3D balanced turbo field-echo sequence. Scanning was performed in an axial plane to include the pons and medulla oblongata, to visualize the cisternal segment of the abducens nerve, by using the following parameters: TR/TE, 6 ms/3 ms; flip angle, 60°; FOV, 176 × 220 mm; matrix, 282 × 352; section thickness, 1.4 mm (0.7-mm overlap with adjacent section); 50 sections; sense factor, 2; scan time, 3 minutes 28 seconds. Reconstruction was performed with zero filling in all three directions, which yielded a voxel size of 0.43 × 0.43 × 0.7 mm.

On MR images, the entire paths of the right and left abducens nerves were traced from the level of the upper medulla oblongata to the level of the upper pons (Fig 1). If the entire cisternal segment of the nerve was not identified, we considered it absent. If the nerve was identified within a limited length from the cisternal segment, we considered it an undetermined case.

Results

The entire cisternal segment of the right and left abducens nerves was well identified in all 10 control subjects. In the first patient, with abduction deficit of the right eye, the right abducens nerve was not identified (Fig 2). In the second patient, with abduction deficits of both eyes, right and left abducens nerves were not identified (Fig 3). There was no undetermined case.

Discussion

This study describes three eyes of two children with abduction deficit that seemed to result from DRS. They presented with only abduction deficit and no other characteristic signs of DRS. They underwent MR imaging to verify the presence or absence of the abducens nerve, and the abducens nerve on the affected side was not identified.

The diagnosis of DRS is essentially based on a clinical examination, which is not difficult when patients present clinically with characteristic signs. Some children with DRS, however, present only with

abduction deficit and no other diagnostic signs of DRS, such as globe retraction, upshoot, or downshoot (5, 6). These other diagnostic signs of DRS develop later, because the lateral rectus becomes more inelastic or fibrotic over time (1, 5, 6). The frequency of correct diagnosis of DRS in children with abduction deficit is reported to be 54% by age 14 months and to increase to 87.5% at 35 months (5). Therefore, cases with only abduction deficit can pose a challenge to the clinician in terms of identifying the etiologic cause.

The most common differential diagnoses of abduction deficit in children include DRS, 6th nerve palsy, and infantile esotropia. The differentiation of DRS from 6th nerve palsy is important because the work-up procedures and the surgical managements of these conditions are different and misdiagnosis can have serious consequences (9). Recession of a rectus muscle is more effective and less traumatic than a resection in treating DRS. Transposition procedure and resection of lateral recti, which are effective in 6th nerve palsy, may worsen the upshoots and the globe retraction and may also produce a marked adduction deficit in DRS (5, 9). In addition, if MR imaging in a patient with abduction deficit shows intact abducens nerves, further study to exclude mass lesions as a cause of abduction deficit should be performed.

Only two reports have addressed the MR imaging findings of DRS (7, 8). One report of one case revealed the absence of the abducens nerve on MR imaging (7), and the other reported that six (54.5%) of 11 eyes (eight cases) with DRS did not have the abducens nerve on the affected side (8). These studies support that a failure to visualize the entire path of the abducens nerve on MR imaging in cases of DRS reflect aplasia and hypoplasia of the nerve. Although even high-resolution, thin-section MR imaging cannot perfectly differentiate aplasia from severe hypoplasia of the abducens nerve, nonvisualization of the abducens nerve on thin-section MR image is helpful for clarifying DRS in patients without clinically characteristic diagnostic signs. This study revealed the absence of the abducens nerve in such cases.

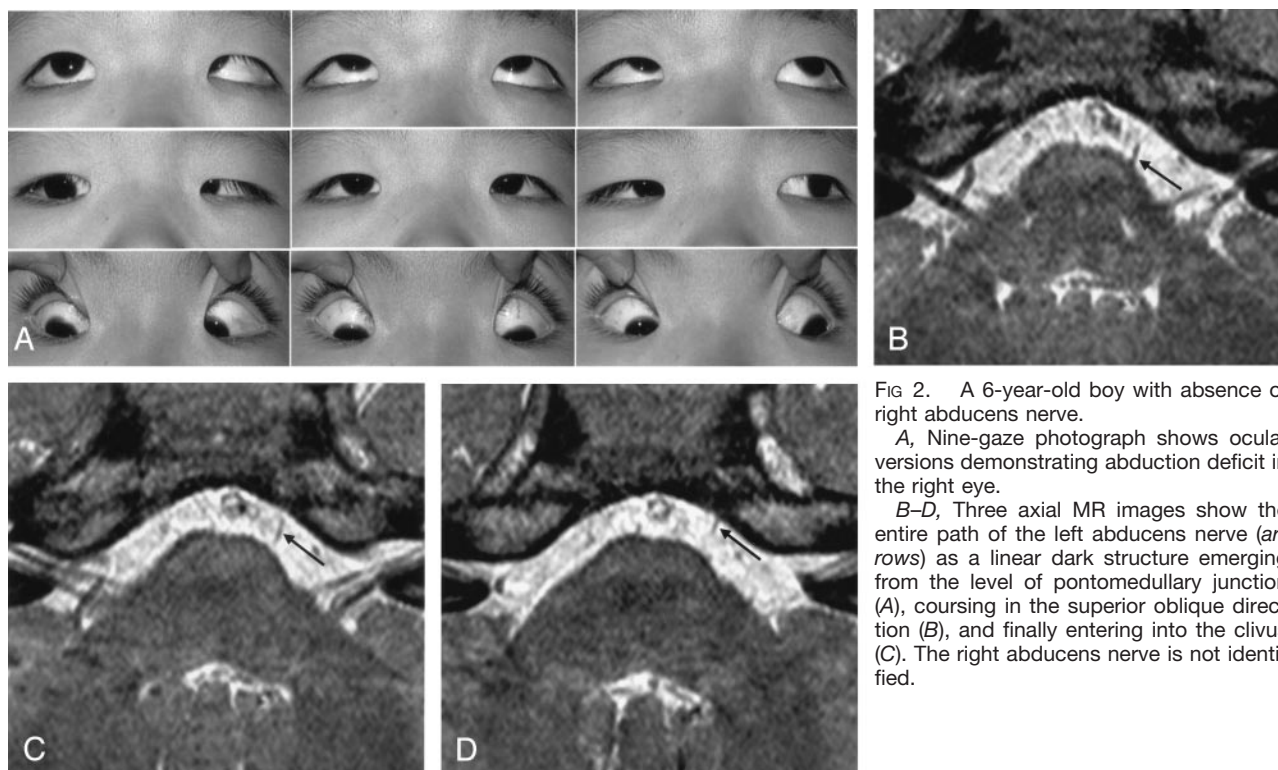


FIG 2. A 6-year-old boy with absence of right abducens nerve.

A, Nine-gaze photograph shows ocular versions demonstrating abduction deficit in the right eye.

B–D, Three axial MR images show the entire path of the left abducens nerve (arrows) as a linear dark structure emerging from the level of pontomedullary junction (A), coursing in the superior oblique direction (B), and finally entering into the clivus (C). The right abducens nerve is not identified.

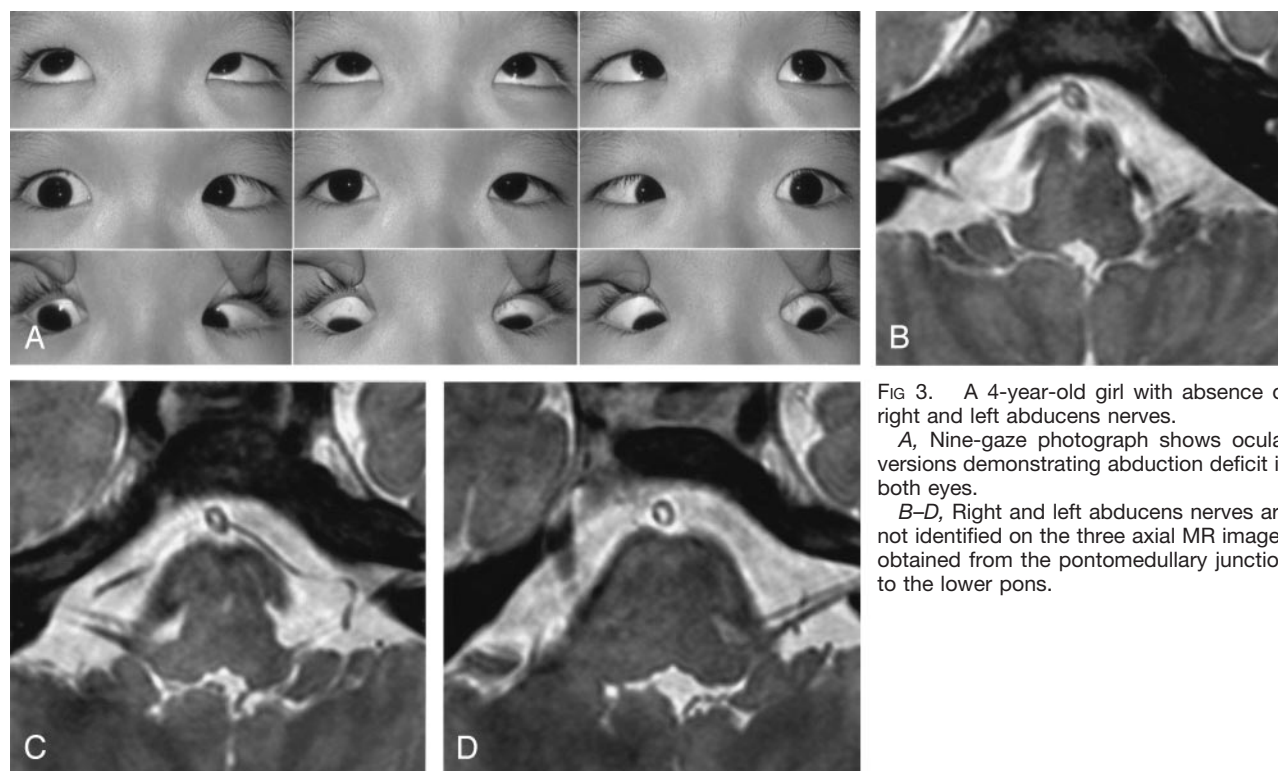


FIG 3. A 4-year-old girl with absence of right and left abducens nerves.

A, Nine-gaze photograph shows ocular versions demonstrating abduction deficit in both eyes.

B–D, Right and left abducens nerves are not identified on the three axial MR images obtained from the pontomedullary junction to the lower pons.

Conclusion

MR imaging is useful for the differential diagnosis of abduction deficit of the eyeball in pediatric pa-

tients. The absence of the abducens nerve strongly suggests DRS in children with abduction deficit. Further detailed studies with a larger number of patients are required.

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