

## Letters to the Editor

### **Torticollis: A Presenting Sign in Idiopathic Intracranial Hypertension**

*To the Editor:* We report a case of spasmodic torticollis as a presenting sign of pseudomotor cerebri, or idiopathic intracranial hypertension. A 32-year-old Saudi female was healthy until three months prior to admission, when she experienced painless, intermittent, involuntary turning of the head to the left. Two weeks later, the deviation became constant and she developed a severe pulsatile, bilateral and retro-orbital headache. She complained of pain in the left occipital and nuchal regions and frequent, transient episodes of blurred vision without diplopia. Six weeks before admission, she lost the vision in her left eye and had dimming of vision in her right eye.

There was no history of fever, trauma, weakness or urinary dysfunction. She complained of numbness in the upper extremities, intermittent tinnitus and dizziness. Similar involuntary turning of the head to the left and neck pain had occurred approximately eight years earlier, when she was 24 years of age. This lasted for six months and she recovered spontaneously.

The patient was moderately overweight with a body mass index of 31. Fundus examination revealed bilateral chronic papilledema with peripapillary hemorrhages and exudates. Visual acuity was 20/400 in the right eye and she could recognize hand movements with the left eye. There was a left relative afferent pupillary defect. Perimetry exam showed a right contracted peripheral field and central scotoma. Her chin turned to the left, with the head tilted to the right and shifted posteriorly; her left shoulder was elevated. On attempting to turn her head to the right, she was unable to reach the midline, and repeated, jerky spasms forced the head back to its original position. The head deviation could be reduced passively past the midline. The torticollis worsened during standing and walking, but partial relief was obtained by resting the head against the wall. The neck posture returned to normal during sleep. On awakening every morning, she was relatively symptom-free for half an hour, during which her torticollis was less severe. The rest of her neurological examination was normal.

Liver and renal functions were normal, as well as hepatitis antibodies, immunoglobulin, cardiolipids, thyroid function, blood count, protein C and S, vasculitis screen, brucella titers, serum prolactin and cortisol levels. Lumbar puncture revealed an opening pressure of 460 mm/H<sub>2</sub>O. Cerebrospinal fluid was acellular and sterile, with normal sugar and protein.

Her torticollis did not improve after the successful placement of a lumboperitoneal shunt. She therefore

required botulinum A toxin injections in the left splenius capitis, left levator scapula, left trapezius and right sternocleidomastoid muscles. She was followed for 12 months, during which time she required multiple botulinum toxin injections; there was moderate improvement.

Torticollis is a focal dystonia characterized by contraction of the neck muscles, resulting in involuntary forced turning of the head. An etiological factor can rarely be identified in cases of focal dystonia, although many secondary causes have been identified. Recently, central or peripheral trauma has been proposed as a cause of dystonia.<sup>2</sup> Our patients fulfilled the criteria for a diagnosis of idiopathic intracranial hypertension (IIH),<sup>3</sup> i.e., increased intracranial pressure with its associated symptoms and signs in an alert and oriented patient, without localizing neurological findings and with normal neurodiagnostic studies.

The close temporal relationship between the onset of torticollis and the development of IIH in our patient raises the suspicion that these two conditions are related, as the torticollis started only days prior to her headache. The other interesting feature of this patient's history was the previous episode of torticollis at age 24 with spontaneous recovery. Spontaneous remission in the course of spasmodic torticollis is more frequent in patients with early onset and these remissions usually occur during the first year.<sup>4</sup>

The neuroimaging studies did not reveal any structural cause for torticollis in our patient. Thus, the clinical picture is more consistent with idiopathic than secondary torticollis,<sup>1</sup> indeed, the previous episode points to a pre-existing predisposition, and the increase in intracranial pressure may have triggered, but not caused, the movement disorder.

An analogy between post-traumatic torticollis and our case is worth considering, as movement disorders may follow relatively trivial trauma.<sup>2</sup> This suggests that a special vulnerability or predisposition to motor disturbances is necessary before involuntary movement is expressed. The exact trigger mechanism of IIH is uncertain; however, the presence of numbness in the arm in our patient, which was relieved by reducing the intracranial pressure, has been reported previously.<sup>5</sup> The neck pain and arm numbness are possibly due to functional stretch in cervical roots, or to dilation of spinal nerve sleeves.<sup>3,5</sup> Both conditions may alter afferent sensory input into the spinal cord or brainstem, leading to reorganization of the local neuronal circuit and enhancement of evoked and spontaneous motor output, possibly mediated by hyperexcitable gamma neurons.<sup>2</sup> In predisposed individuals, this alternation in afferent neuronal input may trigger or accelerate the progression of pre-existing subclinical dystonia.<sup>2</sup>

We conclude that spasmodic torticollis can be a presenting symptom of idiopathic intracranial hypertension providing the individual is vulnerable to motor disturbances. In our patient, this predisposition was possibly related to specific central susceptibility to altered afferent input caused by dilatation of cervical root sleeves, and/or cervical root stretching, due to raised intracranial pressure.

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**Saudi Women's Consent for Invasive Radiological Procedures**

*To the Editor:* There is a recent emphasis on medicolegal aspects of health care practices in Saudi Arabia.<sup>1</sup> A consent is necessary before invasive diagnostic radiologic examinations of interventional procedures. It was made clear long ago that Saudi women can sign their own consent for invasive medical procedures.<sup>2</sup> However, we have noticed some delay in completion of consent forms by hospital staff in a few situations. The objective of this study is to determine the current practice of our hospital staff regarding Saudi females' right to sign consent forms for invasive procedures.

A nonrandom population survey by a questionnaire contained, among other matters, a specific question on whether a Saudi female can sign consent for invasive radiologic procedures. The questionnaire was distributed during grand rounds, meetings or midday clinic. Some meetings were attended by personnel from 22 different

hospitals in Riyadh. The questionnaires were collected at the end of these meetings. This secured a high response.

Demographic information was collected. Results were manually checked for completeness and were subsequently entered on a database file. Data was finally purified. Epistats statistical package was used for analysis.

There were 140 (93%) respondents and 10 (7%) nonrespondents. Five (4%) gave their views by telephone. Ninety-six (69%) were from our hospital and 45 (32%) from 21 hospitals in Riyadh, including teaching, governmental or private hospitals. Fifty-seven (41%) of the 140 respondents were Saudis, 22 (16%) were Western and 60 (43%) were from other countries. One respondent did not declare his nationality. The departments to which respondents belonged are shown in Figure 1. Seventy-seven (55%) of the respondents indicated that a Saudi female can give consent to invasive radiologic procedures, 57 (41%) indicated that they cannot give their own consent, and five (4%) did not know.

Thirteen (22%) of the 57 respondents who said that Saudi females cannot give consent were Saudis, 12 (21%) of those were Western and 32 (56%) were of other nationalities. Thirteen (23%) of the remaining 57 Saudi respondents answered in the negative compared to 12 (54%) of the 22 Western and 32 (53%) of the 60 other nationals.

Abu Aisha has previously refuted the general misconception among the medical profession that a Saudi woman has no right to give consent for essential medical procedures.<sup>2</sup> He showed how such an unfounded conception can be dangerous and explicitly stated the legal standpoint as specified in Article 60 of the hospital management and medical by-laws of the Saudi Ministry of Health regulations supported by Royal decree. Thirty-eight (40%) of the 95 respondents from our hospital deny women their right of consent compared to 19 (33%) of the 45 from other hospitals. This indicates no significant difference in various hospitals in Riyadh. Eighteen (41%) of the 44 female respondents deny women their right of consent compared to 39 (40%) of the 96 males. This

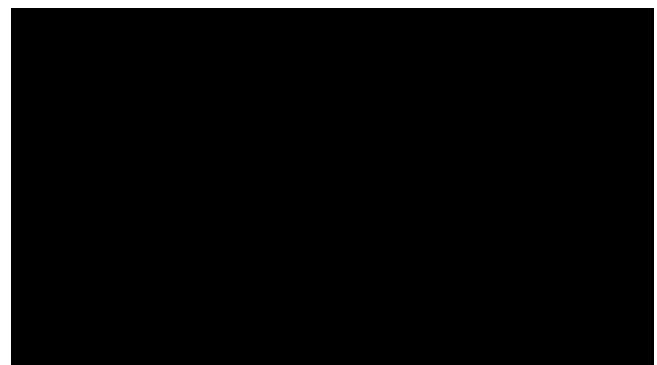


FIGURE 1. Departments of respondents (n=140).

shows that there was no significant difference between male and female respondents in this respect. There was a significant difference between the Saudi and non-Saudi attitude, where only 23% of the Saudis denied females their right of consent compared to 54% of the Western and 53% from other nationalities. This is probably due to prejudice or an overcautious attitude by expatriates.

Despite straightforward legal authorization for mature mentally sound females to give consent for invasive radiologic procedures and previous clarification in the literature, there is still a large proportion of health care providers who would deny females their legitimate right.

This trend is less among Saudi practitioners compared to non-Saudis. Hospital policies and procedures should satisfactorily clarify this matter. Hospital staff should be notified on the legal standpoint.

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### Post-Vaccination Disseminated Bacille Calmette-Guérin (BCG) Infection

*To the Editor:* In-depth investigations of 11 patients at the King Faisal Specialist Hospital and Research Centre who developed disseminated bacille Calmette-Guérin (BCG) infection following routine BCG immunization during infancy,<sup>1</sup> points to severe combined immune deficiency in seven cases. Of the other four, one had isolated T-cell defect, one was healthy, one was diagnosed mucocutaneous candidiasis and one suffered from leukocytoclastic vasculitis. While the demographic and clinical data concerning these 11 patients was succinctly outlined in the article (Table 1 and reference #1), it would be helpful to know more about the batches of BCG vaccines that had been administered to the patients. Like many countries that do not produce BCG vaccine indigenously, Saudi Arabia depends on imported lots for immunization of natives. Three parent strains of BCG, namely Glaxo-1077, Tokyo-172 and Pasteur-1173P2, account for over 90% of BCG vaccines in global use.<sup>2</sup> These differ both in physico-

chemical properties and in mL volume dosage required. The Pasteur strain is administered in 0.1 mL volume doses, while the Tokyo strain vaccine is injected in 0.05 mL volume doses. During a changeover from Pasteur to Tokyo strain vaccine, infants are recorded to have inadvertently been given erroneous double quantities of vaccine.<sup>3</sup> Such episodes are associated with increased frequency of BCG-associated adenitis in the neonatal or early infancy period.

In addition to lifestyle, climate and other demographic factors, the place of domicile and socioeconomic status of the family of the 11 patients<sup>1</sup> would increase dissemination of BCG infection. A change in the BCG strain to be used for vaccination and inappropriate vaccination technique are also of consequence. All of these factors would have to be considered to eliminate further episodes of disseminated BCG infection following national efforts to immunize Saudi neonates with BCG.

Gene probes derived from the insertion sequence, IS986, have been used to distinguish groups of BCG vaccines. Most BCG strains have a single copy of IS986 at the same chromosomal site, while the Brazilian, Japanese and Russian strains have an additional copy at a different, common location.<sup>4</sup> Such gene probes would be useful for characterization of BCG isolates of all patients with disseminated bacille Calmette-Guérin infection retrospectively and prospectively. Epidemiological and molecular biological studies on disseminated BCG in Saudi Arabia<sup>1</sup> would interest numerous developing countries offering BCG vaccines but lacking facilities such as are available in Saudi Arabia. Public health personnel would then be able to tackle similar eventualities in a judicious manner.

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### Reply

*To the Editor:* The issues that have been raised by Dr. Arya in his letter concerning our paper "Clinical Features and Outcome of Eleven Patients with Disseminated Bacille Calmette-Guérin Infection"<sup>1</sup> are appreciated. In regard to the issue of strain of BCG vaccine, the Ministry of Health was using Tokyo-172 until three years ago when it was changed to Pasteur-1173P2 strain. We do not, however, believe that the cases we have reported were related to either change in vaccine, erroneous dose, technique or lifestyle and other environmental factors for the following reasons: 1) The dissemination was not limited to the draining lymph nodes (lymphadenitis). 2) BCG dissemination occurred in patients who received Tokyo and in those who received Pasteur strains alike. 3) It is a well-known fact that patients who are immune compromised are at high risk of developing fatal disseminated BCG. It is extremely rare to have BCG dissemination in an immune competent patient. Therefore, the only valid recommendation not to give BCG vaccine is when there is a family history of primary immunodeficiency, i.e., combined or T-cell deficiency.

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