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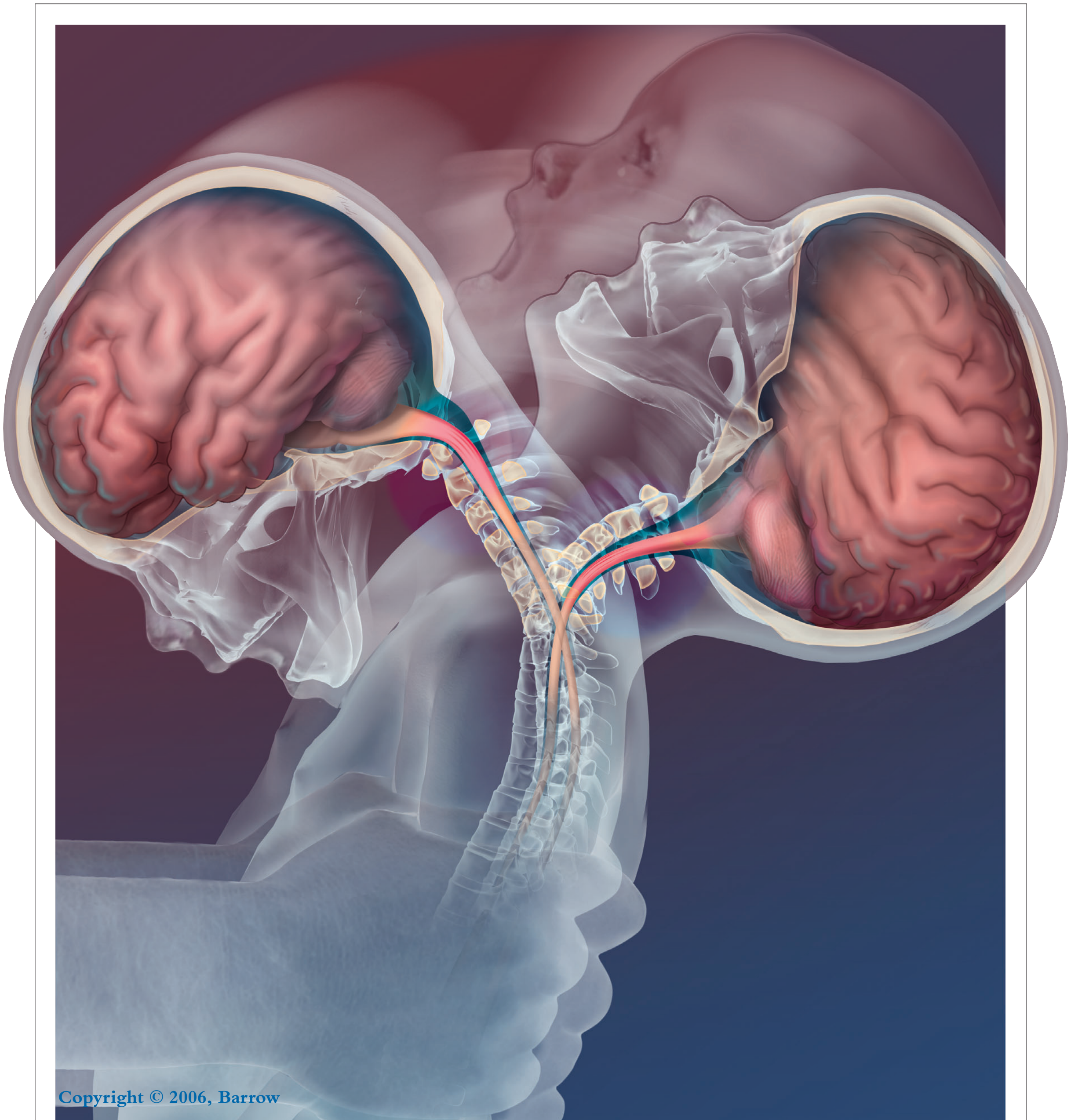

WHIPLASH SHAKEN-BABY SYNDROME WITH SCIWORA

C O M M E N T S

Too often patients cannot avoid their neurological disease or disorder, and medical or neurosurgical treatment may not always offer the hope of a cure. It is no surprise, then, that practitioners in the neurosciences are eager to help prevent unnecessary injury caused by neurological trauma. The case reported by Sana and coworkers represents a particularly disturbing example of avoidable traumatic injury: shaken-baby syndrome. Despite this infant's obvious clinical symptoms, x-rays and computed tomographic (CT) scans showed no evidence of the intracerebral, subdural, or retinal hemorrhage typical of this syndrome. Magnetic resonance (MR) imaging, however, showed abnormalities localized to the cervical spinal cord.

This case exemplifies several important points. First, children with neurological symptoms and no external injuries whose x-rays and CTs appear normal should undergo MR imaging to rule out the possibility of unreported traumatic injury. Furthermore, in such cases, clinicians need to maintain a high level of suspicion for child abuse. With the almost 400% increase in the incidence of SCIWORA (spinal cord injury without radiographic abnormality) in the last two decades, clinical vigilance must remain high to insure early detection and appropriate treatment, and, perhaps, to prevent recurrences. Finally, the findings on MR imaging in this case reinforce the point that the term SCIWORA has become a misnomer: Soft-tissue injuries that do not appear on x-rays and CTs are readily apparent on MR imaging. The term predates the widespread availability of MR imaging and may need to be retired in much the same way that 'occult' cerebral vascular malformation is no longer used to describe a cavernous malformation (ah, but I date myself!).

In another unusual case, imaging showed a brightly enhancing mass in an 83-year-old woman. Such findings in the elderly usually are diagnostic of metastatic disease and prognostic of a poor outcome. Happily, in this patient, the diagnosis was a rare cerebellar hemangioblastoma, which was resected, and the patient's postoperative course was benign. Readers will also find a report of the first auditory brainstem implant performed at our institution. This technology holds great promise for restoring meaningful hearing in appropriately selected candidates, and Barrow is pleased to be offering this state-of-the-art technology. All of these patients received the outstanding care for which Barrow is known, and an integral part of that care is specialized neuroscience nursing. The article on positioning patients with neurological conditions will serve as an excellent reference for all involved in the care of this complex patient population.

We hope that you enjoy these articles and will consider using the enclosed self-addressed stamped envelope to make a tax-deductible donation that will help us continue to share our unusual clinical findings with the medical community. Thank you.

Robert F. Spetzler, MD
Editor-in-Chief

This issue's cover depicts one mechanism of injury when an infant is shaken violently. The incompletely ossified vertebrae in an infant allow a greater range of motion and damage to the spinal cord and nerve rootlets. See the article by Sana et al. on page 4. The illustration is by Mark Schornak and Michael Hickman. The concept for the art is based on a drawing by Steve Harrison.

Whiplash Shaken-Baby Syndrome Causing Cervical Cord Injury Without an Identifiable Radiographic Abnormality: A Case of SCIWORA

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Victims of child abuse typically reach medical attention with visible physical signs of injury. In shaken-baby syndrome; however, clinical findings often reflect neurological deficits while plain radiography or CT may appear normal. Infants who sustain neurological impairment despite normal radiographic and CT findings must be evaluated with MR imaging to rule out a cervical spinal cord injury.

Key Words: child abuse, injury, SCIWORA, spinal cord, trauma, whiplash shaken-baby syndrome

Abbreviations Used: CT, computed tomography; MR, magnetic resonance; SCIWORA, spinal cord injury without radiographic abnormality

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SCIWORA can be seen in pediatric patients who have sustained direct impact injuries. SCIWORA is most often evident in the cervical spine because the hypermobility of the region allows excessive flexion and extension.[2] Direct spinal trauma caused by accidental or nonaccidental trauma is usually accompanied by other abnormalities seen on plain radiography or CT. There are cases, however, in which patients have neurological deficits despite negative radiographs and CT scans; thus, MR imaging is critical to rule out the presence of a spinal cord injury. We present the case of an abused infant who had no intracranial damage but who did sustain cervical damage, a condition known as whiplash shaken-baby syndrome. In this case the patient's plain radiographs and intracranial CT scans were normal, but his cervical MR imaging study was abnormal.

Case Report

An 8-month-old boy, with no significant medical history, was brought to the emergency department for the sudden onset of bilateral upper extremity paralysis and dyspnea. His parents stated that the child was irritable, weak, and breathing rapidly.

On examination, the boy was in mild respiratory distress and appeared lethargic. He was normocephalic and atraumatic. Neurologically, his pupils were equal, round, and reactive to light and accommodation. He had no upper extremity movement and did not withdraw to painful stimulation. His lower extremity function was within normal limits. The remainder of his neurologic examination was unremarkable.

His white blood cell count and the level of his electrolytes were normal. A lumbar puncture was performed. Approximately 15 cc of clear fluid was sent for laboratory analysis. The sample showed a significant elevation of his white blood cell count but was gram-stain negative.

Anteroposterior and lateral cervical spine radiographs showed no fractures or subluxation (Fig. 1). A fluoroscopic examination showed no evidence of instability. CT of the head was negative for intracranial hemorrhage, fractures, or soft tissue abnormalities (Fig. 2). T2-weighted MR images of the cervical and thoracic spine showed abnormal signal intensity within the cervical cord (Fig. 3).

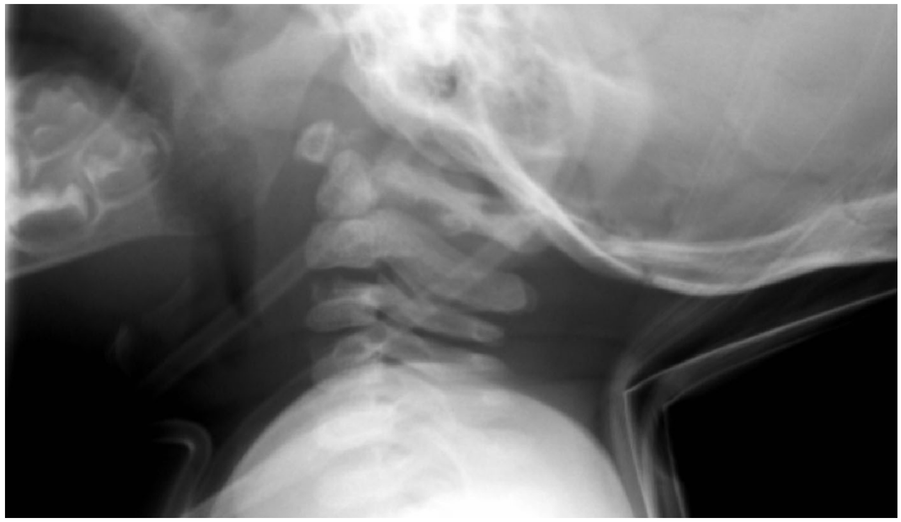

Figure 1. Plain lateral radiograph of the cervical spine of an 8-month-old boy with mild respiratory distress is normal.

Discussion

In the past 20 years, the incidence of SCIWORA as a result of child abuse has increased 400%.[5] Each year the number of estimated cases ranges from 600 to 1400. The incidence of SCIWORA is highest in young patients. In fact, most cases of SCIWORA involve infants before their first birthday. The mean age of patients ranges between 3 and 8 months old.

SCIWORA accounts for about 63.1% of spinal injuries in children 0 to 9 years old and for about 19.7% of the injuries in children between the ages of 10 and 17 years.[5] SCIWORA often involves the cervicothoracic junction.

Figure 2. Axial noncontrast head CT shows no intracranial hemorrhage.

Figure 3. Sagittal T2-weighted MR image shows abnormal signal intensity and swelling extending from the cervicomedullary junction through T-2.

Mechanism of Injury

Extension is often the primary cause of SCIWORA. It is most often caused by a direct impact sustained in a motor vehicle accident or by a fall from a height.[2] In contrast, victims of shaken-baby syndrome experience recurring flexion and extension of the cervical region, a motion that is immensely aggravating to the spinal cord.[6]

The severity of SCIWORA correlates with age and is related to the elasticity and hypermobility of the immature spine. Compared to older children, younger children are less likely to sustain osseous spinal trauma such as fractures. However, when young children do sustain spinal trauma, their immature spinal architecture increases their vulnerability to ligamentous injury. Because the vertebral bodies do not mature until children reach 8 or 9 years of age, severe shaking that causes forceful and repetitive flexion and extension of the cervical vertebrae places almost direct force on the spinal cord. The mismatch between the elasticity of the vertebral column and the spinal cord[4] creates an opportunity for direct injury or for movement of the spinal cord. A child's spine allows as much as 2 cm of stretch whereas the spinal cord itself can withstand less than 0.25 cm of stretch before disruption ensues.[7]

Diagnosis

Shaken-baby syndrome is characterized by retinal hemorrhage, subdural and/or subarachnoid hemorrhage, and minimal or no signs of external craniofacial trauma.[1] Our patient had severe neurologic impairment without evidence of intracranial or external craniofacial trauma on radiographs or CT.

Two criteria are fundamental to the diagnosis of shaken-baby syndrome: (1) a documented history of shaking of the infant and (2) no radiographic evidence of craniofacial trauma. Whether whiplash shaken-baby syndrome can occur without intracranial injury is debated. Authors have argued that traumatic cranial impact is a key component of whiplash shaken-baby syndrome. Our patient is interesting because he not only lacked an intracranial injury, he also lacked an identifiable radiographic abnormality on admission. Thus, the case also fits the category of SCIWORA.

Clinically, the patient had been in mild respiratory distress, which occurs in about 9% of patients with spinal trauma.[3] The child's bilateral upper extremity paralysis and the location of the lesion in the central spinal cord suggested that a central cord syndrome was responsible for the bilateral regions of motor and sensory loss.

MR imaging is the modality of choice for patients suspected of having SCIWORA. It allows evaluation of the spinal cord, ligaments, and discs—structures that cannot be evaluated adequately by radiography and CT. When no abnormalities or intracranial findings are apparent on plain radiographs and CT scans, MR imaging evaluation of the cervical cord may show abnormalities.

Conclusion

Our case demonstrates that children with traumatized necks, despite normal radiographic findings on plain radiographic films and without evidence of intracranial abnormalities, need further evaluation with MR imaging to rule out spinal cord injury suspected on clinical grounds. Given that early detection and care of this syndrome are vital to an optimistic prognosis, health-care teams must consider infant shaken-baby syndrome and SCIWORA in a child with clinical evidence of neurological pathology.

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Case Report: Hemangioblastoma of the Cerebellum in an Octogenarian

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Robert Evans, PA†

In the elderly, symptomatic posterior fossa lesions are usually suspected to be metastatic disease. In this report, we describe the rare presentation of a symptomatic posterior fossa hemangioblastoma in an 83-year-old woman. Familiarity with the classic radiological and clinical features allows clinicians to diagnose these lesions correctly, even in the unusual case of an elderly patient.

Key Words: cerebellum, elderly, hemangioblastoma

Abbreviations Used: CT, computed tomography; MR, magnetic resonance; PICA, posterior inferior cerebellar artery; SCA, superior cerebellar artery

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The differential diagnosis for a homogenously enhancing posterior fossa lesion in the elderly population is relatively limited. Most clinicians suspect metastatic disease unless proven otherwise. We report a rare case of a symptomatic cerebellar hemangioblastoma in an octogenarian.

Clinical Presentation

Over several months, an 83-year-old woman had experienced progressive confusion, memory deficits, and gait difficulties. During this time, the possibility of dementia was raised. At admission the patient suffered from severe nausea, vomiting, confusion, and headache. Laboratory studies revealed a hematocrit of 53%. On examination, she demonstrated lateral nystagmus, left-sided dysmetria, and dysdiadochokinesia. Noncontrast head CT revealed significant transependymal edema and obstructive hydrocephalus (Fig. 1). A large left cerebellar mass was identified. CT of the chest, abdomen, and pelvis showed an adrenal cyst (negative needle biopsy) and no evidence of a primary malignancy. At this time, a bedside ventriculostomy was placed, with an opening pressure of 18 mm Hg. After the procedure the patient's acute nausea and vomiting subsided.

MR imaging showed a brightly contrast-enhancing tumor of the left cerebellum, extending along the ipsilateral tonsil, which herniated below the foramen magnum. Two large peritumoral cysts were also evident. Mass effect on the adjacent brainstem and fourth ventricle produced obstructive hydrocephalus (Fig. 2). Due to the radiographic

Figure 1. Nonenhanced axial head CT shows significant hydrocephalus associated with transependymal edema.

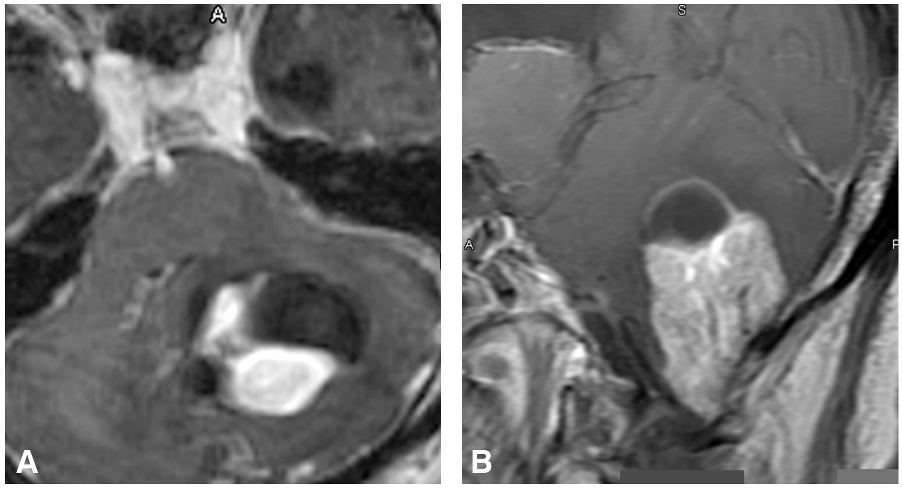

Figure 2. (*A*) Axial and (*B*) sagittal gadolinium-enhanced MR images show a large, brightly enhancing mass in the left cerebellar hemisphere extending down below the foramen magnum. Peritumoral cysts are evident. Note the presence of obstructive hydrocephalus.

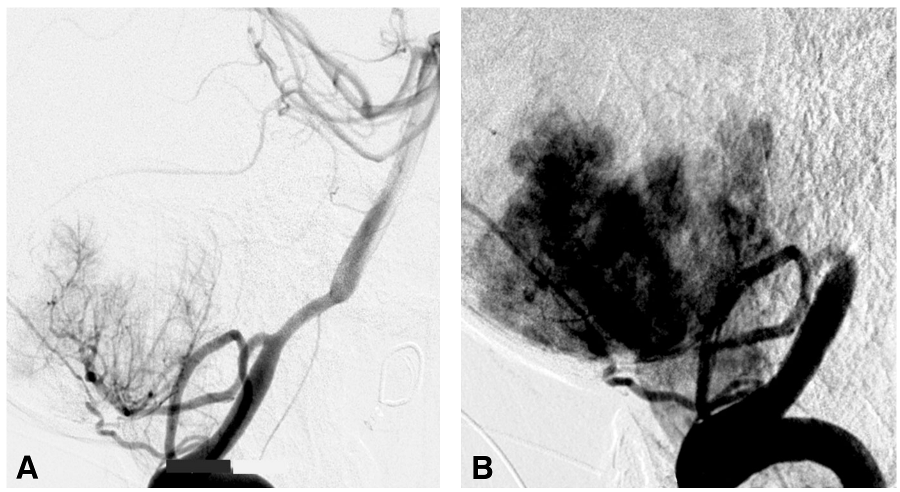

Figure 3. (*A*) Early arterial phase of this left vertebral angiogram shows a large feeding branch from the PICA. (*B*) Capillary phase of the same angiogram shows a dense vascular blush of the tumor mass consistent with, but not pathognomonic for, hemangioblastoma.

features, polycythemia, and clinical history, hemangioblastoma was the leading diagnosis.

The patient underwent cerebral angiography and an attempt at tumor embolization. However, due to the patient's severely tortuous arterial anatomy along the left subclavian and vertebral arteries, embolization could not be performed. Diagnostic imaging indicated that the primary feeding arteries were from the PICA and SCA (Fig. 3A). Angiographically, the arterial blush demonstrated the extraordinary vascular nature of this tumor (Fig. 3B).

Given the radiographic mass effect and dramatic clinical symptoms, the patient underwent a far-lateral craniotomy and gross total resection of the lesion (Fig. 4A and B). Histological analysis confirmed the diagnosis of hemangioblastoma. Except for her baseline cerebellar signs, the patient's postoperative course was benign. Postoperative imaging of the spinal cord revealed no further lesions.

Discussion

The differential diagnosis for an intraaxial enhancing mass of the posterior fossa in the elderly is a metastasis until proven otherwise. This diagnosis corresponds with a poor overall prognosis. However, as this case demonstrates, clinicians should be aware of hemangioblastomas as a rare cause of brightly enhancing posterior fossa masses in the elderly.[2] Features such as intense enhancement, flow voids, and peritumoral cysts can be radiographic indicators of hemangioblastomas. Extremely vascular metastatic tumors, such as renal cell carcinoma hypernephroma, can mimic hemangioblastomas radiographically.[7] Therefore, it is reasonable to rule out a primary carcinoma as a potential etiology in the elderly or in patients with risk factors for cancer. Other rare causes of intraaxial tumors in the posterior fossa include glioblastomas, ependymomas, choroid plexus carcinomas, medulloblastomas, and lymphomas.[11]

Hemangioblastomas are benign vascular tumors. If suspected clinically, aggressive treatment may be warranted based on the patient's clinical symptoms and radiographic evidence of mass effect. Surgical management is based on a case-by-case analysis. Preoperative embolization, while not mandatory, can be useful for large tumors and for lesions adjacent to the brainstem or fourth ventricle.[9] Surgical extirpation is curative. Radiosurgery, although supported by

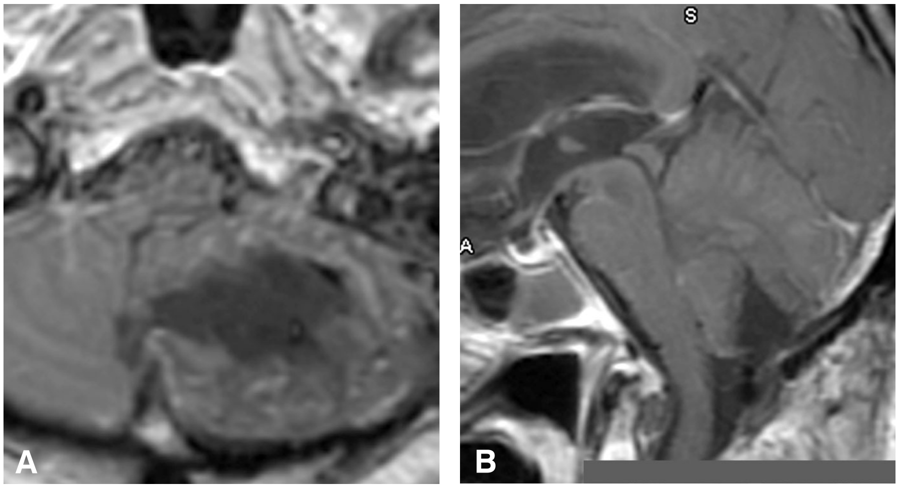

Figure 4. (*A*) Axial and (*B*) sagittal gadolinium-enhanced MR images confirm gross total resection of the tumor. Frozen and permanent sections (*not shown*) confirmed the diagnosis of hemangioblastoma.

some authors,[5,8] has no proven role in the treatment of this disease.

Hemangioblastomas compose 1 to 2.5% of all intracranial tumors, with about 85% occurring in the cerebellum. Less commonly, they are found in the spinal cord, medulla, and cerebrum.[3,6,12] Histologically, hemangioblastomas are composed of three cell types: pericytes, endothelial cells, and stromal cells.[4] Due to the vascular nature of these tumors, histopathological sections frequently catch branched arterioles in longitudinal sections, creating the classic "staghorn" feature. Approximately 60% of hemangioblastomas are solid tumors, while 40% can have cystic components. Cysts contain clear, straw-colored fluid, and the cyst wall does not need to be removed during resection. Enhancing mural nodules and solid components, however, must be treated with gross total resection.

Hemangioblastomas usually become symptomatic in the third to fourth decades of life. Polycythemia can occur in 10 to 20% of patients. Eighty percent are sporadic, and 20% can be associated with von Hippel-Lindau syndrome. The latter is characterized by an autosomal dominant heritance pattern; multiple hemangioblastomas; and other systemic tumors such as renal cysts, renal cell carcinomas, pheochromocytomas, and retinal angiomatoses.[1]

The advanced age of our patient is noteworthy. This case report demonstrates that a hemangioblastoma can be a rare cause of a primary intraaxial tumor in the cerebellum in elderly individuals. Given the classic appearance of these lesions on MR imaging and angiography and the absence of any primary source of tumor, the diagnosis of hemangioblastoma was correctly suspected preoperatively. Because the patient was symptomatic from both mass effect and cerebellar signs, we believed that surgical resection was a reasonable option in this otherwise healthy octogenarian.[2,10]

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The First ABI at the BNI: Case Report and Literature Review

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First introduced in 1979 by Drs. House and Hitselberger, auditory brain stem implants are capable of providing useful hearing for a patient suffering from bilateral retrocochlear deafness. The initial design of the ABI featured a single-channel ball-type electrode. ABIs have since evolved to include between 8 and 21 electrodes. In the year 2000, the Food and Drug Administration approved a multichanneled ABI for the treatment of neurofibromatosis type 2. Currently, more than 400 ABIs have been implanted throughout the world, including in the United States, Australia, Germany, Italy, the United Kingdom, Poland, and Croatia. Sound-only open-sets speech comprehension has been reported in a minority of patients who have received ABIs, but most patients derive maximum benefit from using the implants as an adjunct to lip reading. In this regard, ABIs perform as well as single-channel cochlear implants, on which their design has been based. We present our initial experience with ABIs and review the literature.

Key Words: acoustic neuroma, auditory brainstem implant, auditory prosthesis, neurofibromatosis type 2, vestibular schwannoma

Abbreviations Used: ABI, auditory brainstem implant; BCI, brain-computer interface; CT, computed tomography; FDA, Food and Drug Administration; MR, magnetic resonance; NF 2, neurofibromatosis type 2

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Much of the emphasis in current research on BCI technology is on restoration of neurological function. BCI technologies intended to extend human capabilities may have appeal for military applications. Thus far, however, all available examples of this technology have been developed with the goal of ameliorating the symptoms of disease, such as implantation of deep brain stimulation electrodes for the treatment of Parkinson's disease.

Of special interest is the potential for these devices to restore special sensory modalities, in particular, vision and hearing. Efforts to obtain viable prosthetic vision are still in their infancy.[6] In contrast, ABIs are available technology that provides prosthetic auditory information directly to the cochlear nucleus at the pontomedullary junction level of the brain stem. Specifically, in the year 2000, the FDA approved multichannel ABIs for use in patients with the diagnosis of neurofibromatosis type 2 (NF 2) who are 12 years of age or older and who have reasonable expectations about the device's utility.

At least five models of ABIs, including the Nucleus 24 Contour and Nucleus 22 ABIs (Cochlear Corporation, Englewood, CO), the MXM Digisonic ABI (Laboratoires MXM, Côte d'Azur, France), the Med-El ABI (Med-El Corporation, Research Triangle Park, NC), and the Clarion ABI (Advanced Bionics, Sylmar, CA), are available. To date, no evidence suggests superior performance of any particular model of ABI.

Regardless of the manufacturer, all ABIs share certain common features, including a digital speech processor, transmitter coil, and a receiver-stimulating unit that is connected by cable to an electrode array of 8 to 21 electrodes overlying the cochlear nucleus. The Nucleus 24, the most widely implanted device, features an electrode array of 21 platinum discs embedded in a Dacron mesh. The device is placed within the lateral recess of the fourth ventricle. The electrode is connected by a cable to a receiver stimulating unit, which itself is seated in the occipital region of the skull. An additional wire serves as a ground electrode and is implanted in the temporalis muscle. Therefore, the entire device is subcutaneous and communicates with the digital speech processor through a radio transmitter coil. This second component includes a microphone, which digitizes incoming sound, processes speech, transmits this digital signal through the radio transmitter coil to the receiver stimulator located underneath the scalp, and then transmits it from there to the brain stem.

Given the various number of channels that patients can use to interpret auditory stimuli, the utility of these devices relies on cooperation and training for the successful interpretation of the auditory input. With the Cochlear digital speech processor, the patient can adjust the speech processor through four user-selectable programs and can control the volume and sensitivity.

Indications

The FDA has approved ABIs for patients with NF 2 who are 12 years of age or older and who have reasonable expectations and motivation. NF 2 has an incidence of 1 in 40,000. The phenotype is associated with an autosomal dominant mutation of the merlin gene on chromosome 22, resulting in bilateral vestibular schwannomas. This latter feature is pathognomonic for the diagnosis. Vestibular schwannomas associated with NF 2 are more aggressive or more likely to invade the vestibulocochlear nerve than nonsyndromic schwannomas.

An additional set of characteristics establishes a patient as an optimal candidate for an ABI: good overall state of health, acceptable vision, an interest in spoken communication, and acceptable anatomical features. Vision is important because in terms of speech comprehension, the greatest utility is typically derived from using the ABI as an adjunct to lipreading. Perhaps most importantly, however, patients must exhibit high motivation and have sufficient family support.

Given the largely successful experience with ABI in the setting of NF 2, it is not surprising that the indications for ABI placement have evolved to include other conditions, including unilateral vestibular schwannoma (i.e., nonsyndromic) in an only hearing ear,[2] deafness as a result of bilateral skull base trauma,[1] cochlear nerve aplasia or hypoplasia,[4] and cochlear ossification.[7] In theory, any cause of bilateral retrocochlear deafness may suffice as an indication for implantation of an ABI. To date, however, experience with ABI placement for indications other than NF 2 is limited to Europe. The only absolute contraindication to the placement of a cochlear implant is the presence of active infection.

Case Report

A 56-year-old male was diagnosed with NF 2 at 21 years of age. The patient underwent a translabyrinthine resection of the vestibular schwannoma on the left side 20 years earlier at an outside institution. At the time of his evaluation at our institution, he had experienced progressive hearing loss on the right side. His medical history was significant for insulin-dependent diabetes mellitus, peripheral neuropathy related to the diabetes, and seizure disorder. His current medications included Neurontin, OxyContin, insulin, Skelaxin, omeprazole, and Capitrol. He also complained of difficulties with swallowing and clearing secretions and occasional bouts of urinary and bowel incontinence.

Physical examination revealed myoclonus, dystonic posturing of fingers, weakness of the intrinsic hand muscles, and bilateral foot drop. The patient's vision was excellent. He wore a hearing aid in the right ear and depended on lip reading for communication.

Preoperative audiography demonstrated an elevation of speech awareness threshold to 90 dB and a pure-tone discrimination threshold elevation of 106

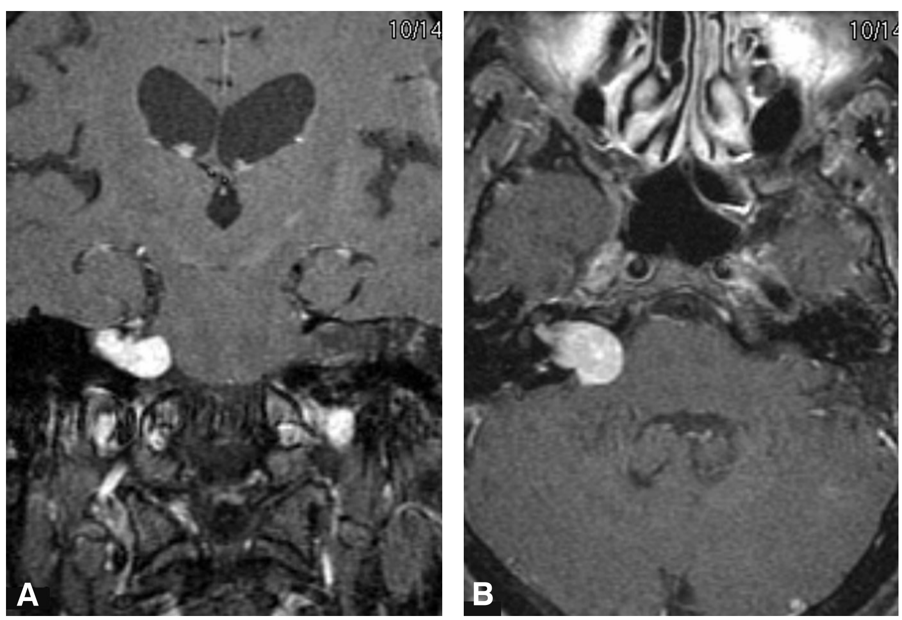
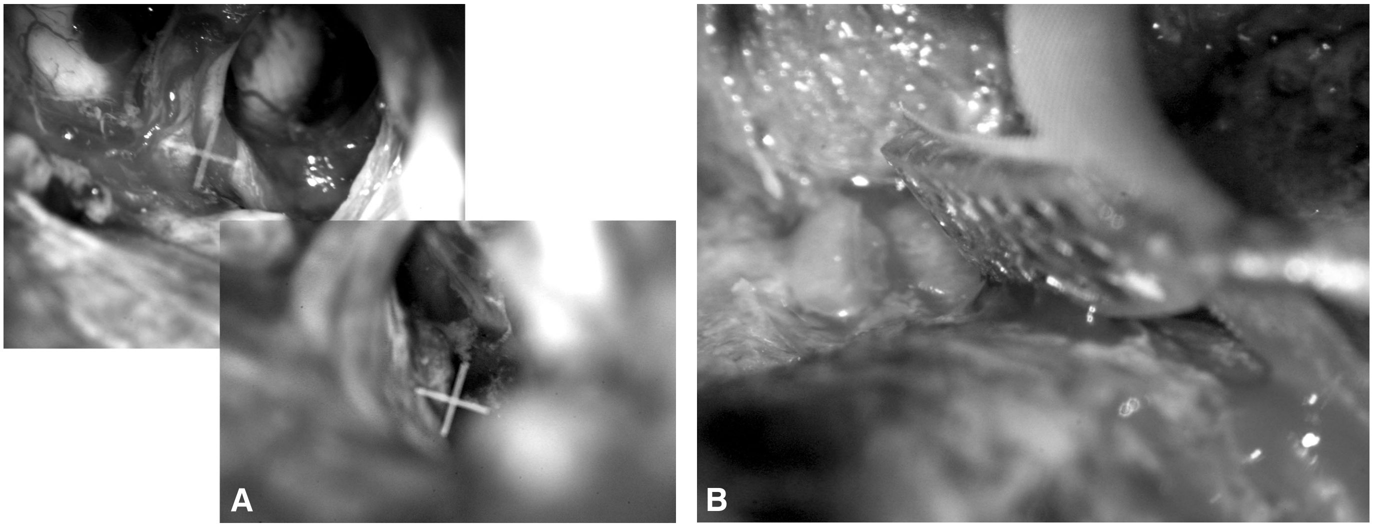


Figure 1. Preoperative (*A*) coronal and (*B*) axial contrast-enhanced MRIs show a homogeneously enhancing tumor of the right cerebellopontine angle with an 'ice-cream cone' profile characteristic of a vestibular schwannoma.



dB. Preoperative imaging (Fig. 1) showed a 13 x 20 x 12 mm acoustic schwannoma on the right side and surgical changes on the left side without evidence of tumor (the site of previous surgery). Comparison with MR images obtained 2 years earlier demonstrated modest tumor growth (previous size 12 x 15 x 10 mm).

The patient was offered the following options: (1) continued observation, (2) radiosurgery, or (3) resection of tumor with anticipated loss of hearing and placement of an ABI. During preoperative counseling, it is important to stress the risks, benefits, and alternatives to surgery. Deafness is likely to be universal in neurofibromatosis, but not all patients are ideal candidates for an ABI. Given that our patient was already anacoustic in the left ear, there was no chance of maintaining useful hearing regardless of treatment modality (i.e., surgery or radiosurgery). We believed that the patient's best chance of retaining useful hearing on the right side after resection of the tumor was an ABI. The patient elected to undergo surgical resection of the tumor and placement of an ABI. Informed consent was obtained.

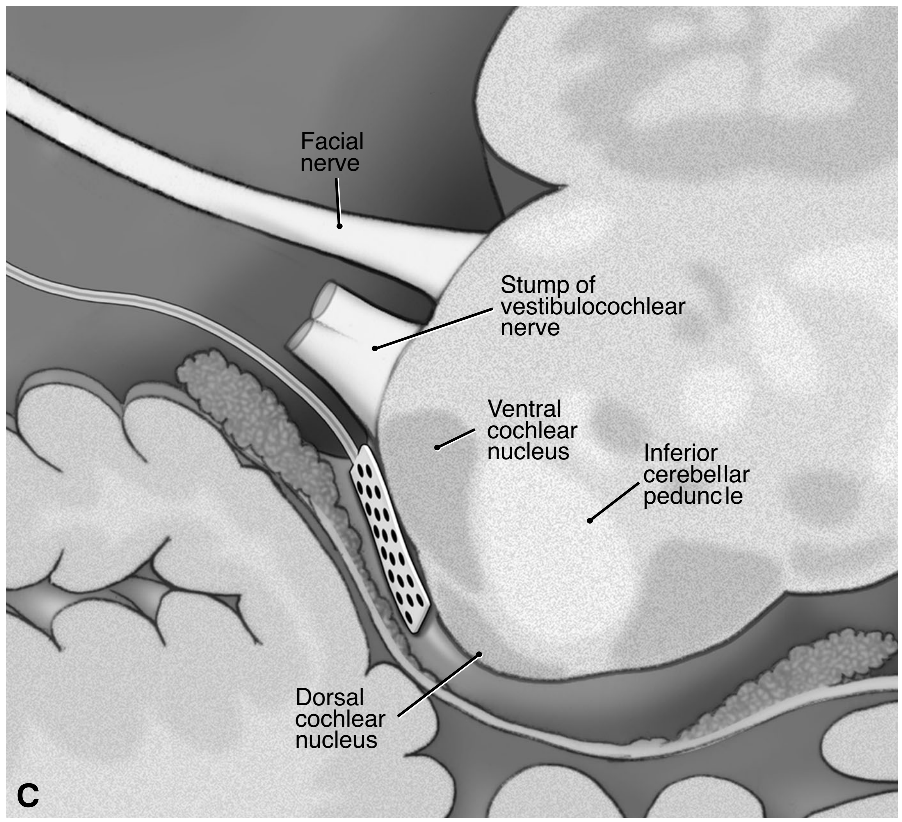


Figure 2. (*A*) Intraoperative photographic montage shows the seventh and eighth cranial nerve complex (*upper crosshairs*) and the ninth cranial nerve (*lower crosshairs*). (*B*) Intraoperative photograph of electrode array being advanced into the lateral recess of the fourth ventricle. (*C*) Schematic illustration of an ABI electrode in the lateral recess of the cochlear nucleus.

The patient was positioned supine with his head turned left to expose the right ear. A standard translabyrinthine approach to the tumor was performed. The tumor was resected in the typical fashion. When resection of the tumor was completed, the facial nerve, the stump of the vestibulocochlear nerve at its entry to the brain stem, and the glossopharyngeal nerve were identified (Fig. 2). The lateral recess of the fourth ventricle was identified by following the tenia of the fourth ventricle and a small tuft of choroid plexus emerging from the foramen of Luschka. The implant was then advanced within the foramen of Luschka along the pontomedullary surface (Fig. 2B).

Despite the presence of significant signal artifact from the ABI (Fig. 3), postoperative MR imaging confirmed

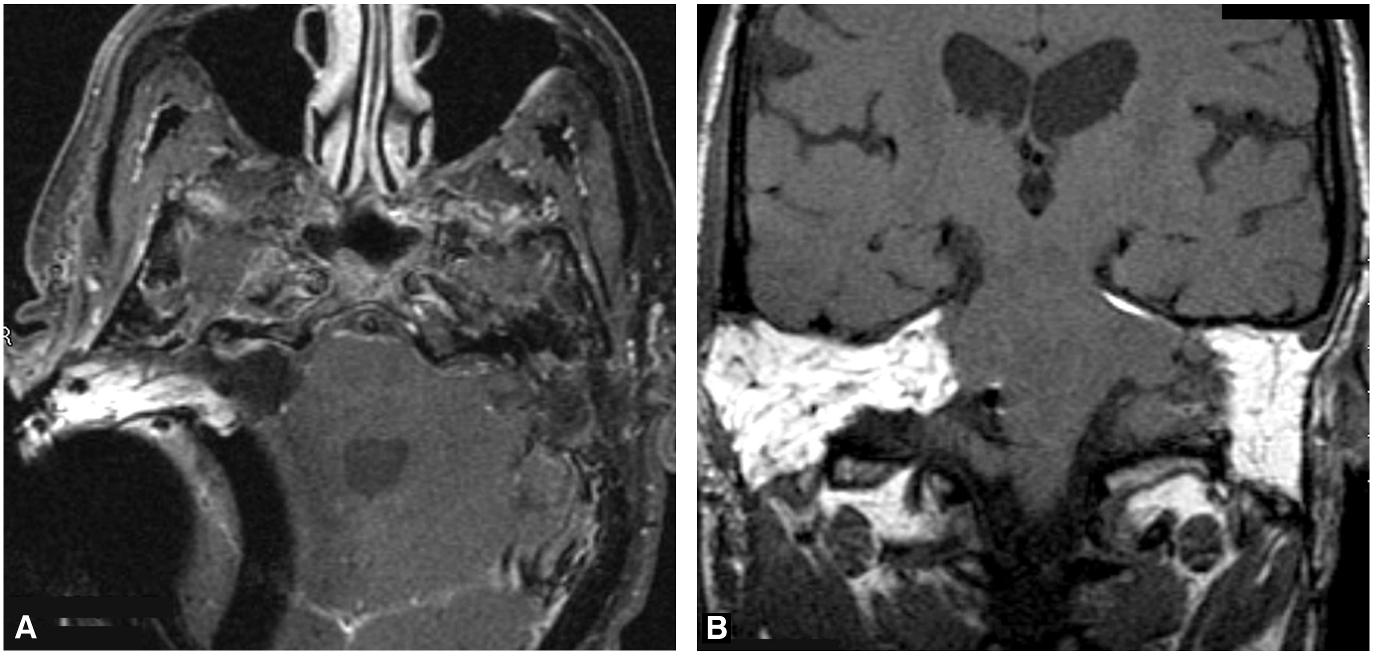


Figure 3. Postoperative (*A*) axial and (*B*) coronal MR images confirm complete removal of the tumor and show the fat packed in the translabyrinthine approach defect. The artifact on the images is caused by the ABI receiver-stimulator.

complete resection of the tumor with fat packed into the translabyrinthine defect. CT demonstrated satisfactory placement of the electrodes (Fig. 4). Pathological analysis confirmed the diagnosis of a typical vestibular schwannoma.

Approximately 8 weeks after surgery, the patient's ABI was turned on. During the initial programming with stimulation of a single channel, he reported the reception of auditory percepts on four channels. At his 15-month follow-up examination, the patient underwent 10 pitch-ranking maps. He is a daily user of his ABI, aware of environmental noises, and able to distinguish his wife's voice.

At the initial programming of the ABI, the patient had no auditory-only channels and auditory percepts were associated with a shock-like sensation. With subsequent programming, this nonauditory percept was eliminated. Formal speech-perception testing, as assessed by the Hearing in Noise Test presented via live voice, revealed speech comprehension rates of 12% (auditory only condition), 16% (visual only), and 40% (auditory + visual). Importantly, both the patient and his wife have noticed dramatic improvement in his hearing when he uses the device and at 24 months follow up were pleased with the results.

Discussion

In the English language literature, more than 180 patients with an ABI have been reported.[3,5,9,10,12] The largest single series is from the United States Clinical Trial,[5] which enrolled 92 patients whose average age at implantation was 33.9 years (range, 12.7 to 67.5 years). In two-thirds of the patients, their ABI was placed at the time of a second surgery. One-third received their ABI upon resection of their initial schwannoma (i.e., in the presence of a hearing ear on the contralateral side). Fifty-nine percent of the patients were female and 41% were male. Two patients died before the initial activation of their implants. Of the 90 remaining patients, 74 received auditory percepts upon stimulation (82.2%).

At the time of initial stimulation, most patients also experienced nonauditory sensations. The most common site for nonauditory percepts included the ipsilateral head and neck followed by the ipsilateral torso and upper and lower extremities (60%, 14%, 10%, and 11%, respectively). Although rare, the rate of contralateral percepts was 6%. Approximately 10% of patients received auditory sensations only, and another 7% received nonauditory percepts only. The origin of nonauditory percepts is attributed to the proximity of distal electrodes to the inferior cerebellar peduncle. In general, the character of nonauditory sensations has typically been described as tingling, dizziness, visual jittering, or muscle twitching.

In terms of improved speech comprehension, the results from the United States Clinical Trial mirror the percentage of patients who received auditory percepts. Eighty percent reported that they benefited from use of the device, and 75% reported wearing the device daily. When used in conjunction with lip reading, the ability of 85% of the patients to understand sentences after 3 to 6 months significantly improved based on the City University of New York (CUNY) score. Twelve percent of patients demonstrated clinically significant open-set sentence recognition in the auditory-only condition.

As recently reviewed,[8] these results have been replicated both domestically

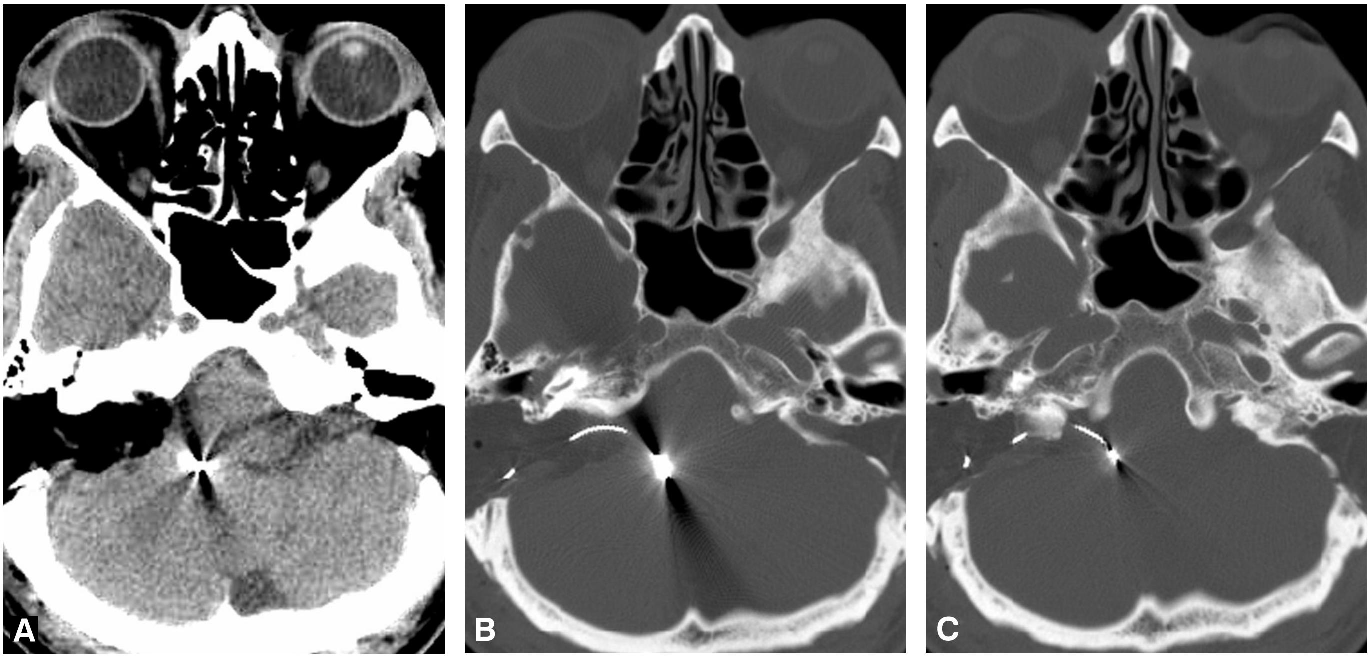


Figure 4. (*A*) Soft-tissue window and (*B*) bone window CTs of the head obtained immediately after surgery and (*C*) on the day of initial stimulation of the ABI confirm that the device did not migrate during the follow-up interval.

and internationally, including by the Digisonic Clinical Trial reported in the journal, *Otology Neurotology,* in 2002[12] and by European centers, including Verona,[3] Hannover,[9] and Freiburg.[10] The percentage of patients receiving auditory sensations ranged from 86% in the Digisonic study to 100% in the Hannover and Verona studies. Between 75 and 100% of patients were reported as daily users (the Digisonic trial and Verona study, respectively). There appears to be no association with the number of electrodes and efficacy of the device. The United States Clinical Trial was performed with the 8-electrode Nucleus 22 model. European data have been reported with either 14- or 21-electrode models. However, the results associated with the different devices have not varied significantly.

Preoperatively, it is important to counsel patients not to be discouraged or disappointed with the unnatural quality of the sound associated with their ABI. The sound has been likened to a muffled loudspeaker. However, the results of audiologic testing have continued to improve as long as 8 years after implantation. With appropriate expectations and patient cooperation, ABIs restore meaningful hearing to most patients. Based on the long-term follow-up from the experience at the House Ear Institute,[11] 87% of patients exhibited scores on closed-set testing of word recognition above the level of chance. Environmental sound discrimination was more than 50%, and sentence recognition scores improved by 26%. Importantly, even when sound-only open-set speech recognition is not obtained, patients benefit from the perception of environmental sound and their ability to read lips improves significantly.

Conclusion

Future designs of ABIs have centered on the development of the penetrating brain stem implant. However, initial results with this implant have been disappointing (Hitselberger W, personal communication, 2005). These results, which may reflect the development of perielectrode scar tissue, may not portend well for the use of other penetrating BCI strategies. Alternatively, the limited performance of the penetrating ABI may reflect a reliance on stimulation of deep cochlear nuclear structures, while providing enhanced tonotopic representation of pitch theoretically may diminish speech-recognition capability. Research into speech-processing algorithms may be another promising avenue for improvement of speech recognition. However, caution must be exercised in the exposure of the brainstem to charge. Hence, the ranges by which these parameters can be safely modulated may be limited.

As our experience with ABIs increases, innovations in electrode design and our improved understanding of plasticity in the ascending auditory pathway will be accompanied by improvments in the performance of these devices. Nevertheless, at present, ABIs truly reflect the state of the art of BCI technology.

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Patient Positioning: More Than Just "Turn Every 2h"

Terry Bachman, RN

For patients with neurological conditions, the routine nursing task of turning and repositioning requires special attention. Neuroscience nurses must select the optimal adjustments for the head of the bed and the best equipment and means of mobilization when caring for this complicated patient population. This article describes the careful balance of intervention and vigilance, hallmarks of neuroscience nursing care, in terms of patient positioning.

Key Words: nursing, positioning

Abbreviations Used: CSF, cerebrospinal fluid; EEG, electroencephalography; ICP, intracranial pressure

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Positioning patients with neurologic injury may seem like a routine nursing task. However, neuroscience nurses must consider many factors when they position patients. Nurses must optimize the neurologic function of patients with a brain injury while protecting them from the hazards associated with immobility. Patient positioning is not just a matter of "turn every 2h" or "head of bed 30 degrees at all times." Delicate balances in the circulation of blood, CSF, and cellular fluid as well as the dynamics of cerebral elastance and anatomical shift must be considered when patients with different types of brain injury are positioned. Important factors related to positioning also affect cognition, peripheral nerve function, joint mobility, pulmonary function, and general recovery from brain injury.

Position of Head of Bed

Once a cervical injury has been ruled out, several factors determine how the head of bed is positioned in patients with brain injuries. Research by physicians and nurses offers conflicting findings regarding the best position for the head of the bed for patients with neurologic injury. Positioning the head of the bed at 30 degrees is associated with lower ICP compared to a flat or lower elevation (Fig. 1).[6] Others advocate individualizing this decision for patients with large hemispheric strokes.[11] Elevating the head of the bed to 30 degrees decreases ICP, but it also decreases systolic arterial blood pressure. The effects of position change on other variables, including measures of cerebral perfusion pressure, jugular venous oxygen saturation, and velocity of cerebral blood flow, have also been studied.[6] Most

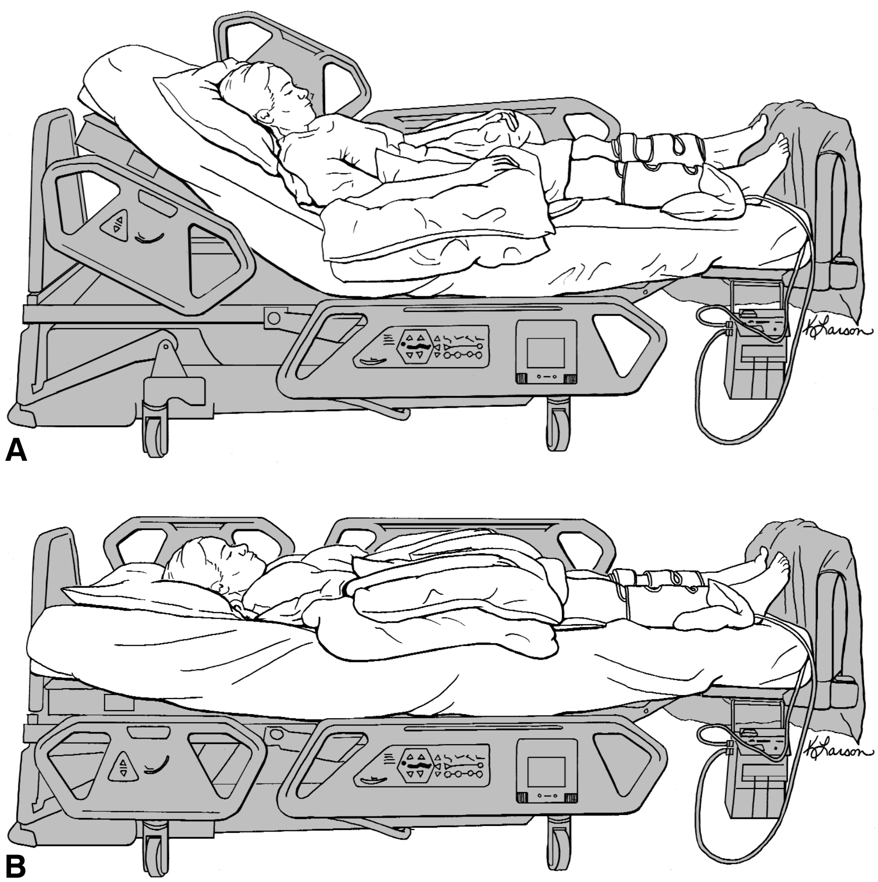

Figure 1. (*A*) The head of the bed is positioned at a 30-degree angle compared to a (*B*) horizontal or flat position.

researchers advocate raising the head of the bed to 30 degrees for patients with elevated ICP.[6] Researchers also advocate elevating the head of the bed to augment venous jugular return, arguing that the benefits outweigh the effects of lowered systolic arterial blood pressure.[8]

The position of the head of the bed may be individualized for patients with a subdural hematoma. Patients with chronic subdural hematomas tend to be from an older population and therefore have diminished cerebral elastance and some degree of brain atrophy. At our institution, such patients are managed with the head of bed flat after the subdural hematoma has been evacuated. This position prevents gravity from pulling the brain away from the dura. Theoretically, the flat position allows the CSF to float the brain in a more neutral position. Once the cerebral tissue has had a chance to expand to fill the void of the evacuated area, the head of the bed is elevated as tolerated (as determined by the physician's evaluation of a patient's routine computed tomography scans).

Patients with subarachnoid hemorrhages, cerebral contusions, epidural hematomas, and cerebral infarcts need the head of the bed positioned at 30 degrees. The head itself is positioned neutrally with the nose, chin, and sternum in alignment (Fig. 2). This position optimizes jugular venous drainage and decreases the chance of aspiration related to craniopharyngeal weakness. The head-up and neutral positions are maintained while the patient is alternated through the supine, right, and left side-lying positions. Pillows and rolled towels are placed strategically to prevent gravity or hyperactivity from placing paralyzed or overly active limbs, respectively, in a position that decreases venous return (Fig. 3).

For patients who have undergone transsphenoidal resection of a pituitary tumor, the head of bed is maintained at more than a 45-degree elevation to promote venous drainage of the head and brain. The drainage afforded by this position helps decrease swelling of the soft tissues of the face and helps to decrease fluid pressure on the surgical site. This elevation is instituted during the early postanesthesia phase while nurses team together to lift and position the patient in bed. It is maintained when the patient is alert and moving independently.

Determining the best position for patients with a CSF leak is another challenge. At our institution, the neuroscience nurses reason through the best management for each patient in collaboration with the physicians. In general, the head of the bed is elevated at all times for patients with rhinorrhea and otorrhea. Patients with leakage from spinal incisions are initially maintained with the head of the bed flat. If the patient has a lumbar drain inserted to manage the leak, stipulations for positioning may include gradually increased activity with titration of the height of the drainage chamber to achieve a desired volume of drainage.[10] Activity may be increased to include sitting in a chair, assisted ambulation, or both.

While the patient is standing and walking, the nurse closes the drain. When the patient is seated and comfortably stationary, the nurse levels and reopens the drain. During all repositioning, the nurse maintains the integrity of the drainage system. Pulling or tugging the drainage tube is avoided. The tube is observed carefully to avoid over- or underdrainage.

Severe Intracranial Hypertension

Patients with severe intracranial hypertension requiring treatment with a pentobarbital coma must be maintained in a neutral head position (Fig. 2) to optimize pulmonary function. Pentobarbital induces a comatose state, creates total flaccidity of the muscles, and interrupts basic reflexes. The patient's natural defenses against aspiration, atelec-

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Figure 2. (*A*) Patient shown in optimal neutral alignment. (*B*) Misalignment can compromise a patient's jugular venous drainage and should be avoided.

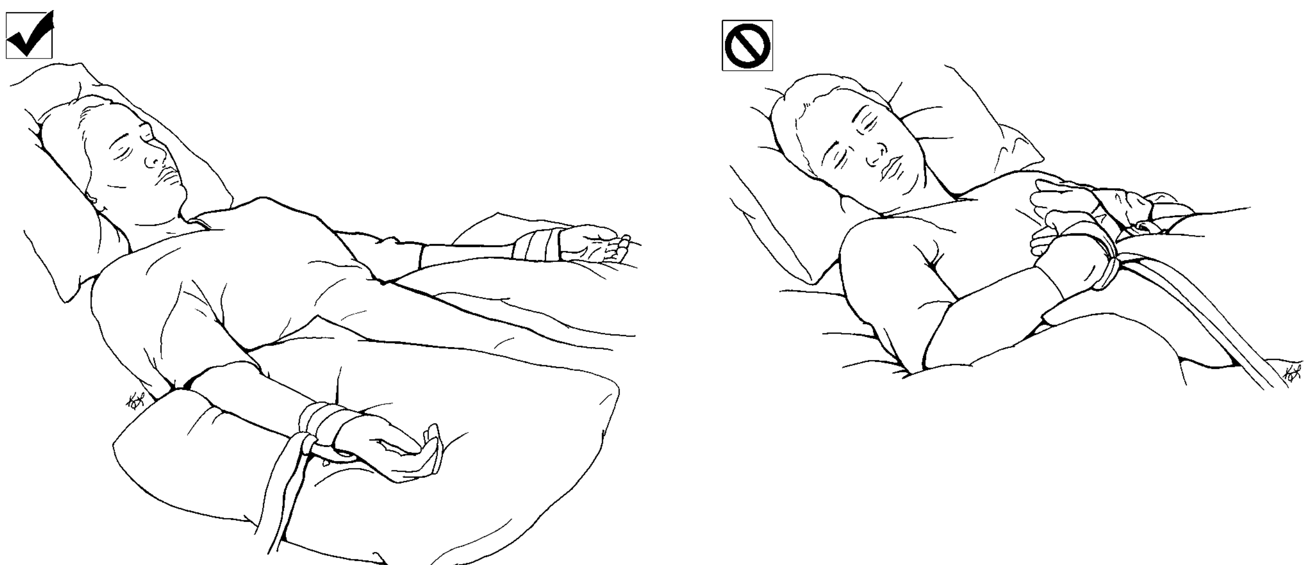

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Figure 3. (*A*) Pillows positioned to elevate the arms in a supine position at the level of the heart to improve venous return and to minimize the risk of injury to the ulnar nerve. (*B*) Hyperflexed arms, which can impede venous return and decrease mobility, should be avoided.

tasis, and immobility are thereby paralyzed. Patients in this induced comatose state may experience profound increases in ICP merely from the stimulation associated with repositioning.

A solution to the complications associated with patient immobility has been the use of rotational beds. These beds use a motorized mechanism to turn patients from supine to side-lying positions at frequent intervals while eliminating the stimulation that can increase ICP. The head and limbs are maintained in controlled neutral positions via bolsters adjusted by the nurse according to the contour of an individual patient's habitus. The specialized beds provide the rotation necessary to mobilize pulmonary secretions and to promote pulmonary circulation.

Several types of rotational beds are available, and each has its own advantages and disadvantages. At our institution Roto Rest® beds (Kinetic Concepts Inc., San Antonio, TX) are preferred (Fig. 4). The firm bolsters on this bed achieve neutral alignment and continue to maintain the patient's position after hours of rotation. With this bed, the patient requires less frequent repositioning by the nurse.

The air pillow-styles of other rotational beds provide support and rotation but do not seem to adequately maintain neutrality of the axis formed by the head, chin, shoulder, and sternum, which optimizes jugular venous return. Air-style beds, however, relieve pressure and provide maximum degrees of rotation. Consequently, attention to skin integrity, especially over the bony prominences during activities of daily living, must be vigilant. Prolonged supine positioning can cause a decubitus ulcer to form on the occiput. To maintain skin integrity, waffle air products can be used to cushion the occiput.

Even with the use of the best rotational bed, further intervention via manual positioning is sometimes needed to manage high ICP. Attention to subtle differences in a patient's physiologic responses to turning may show that a patient's ICP is higher when rotated to one side compared to the other. Patients may be intolerant to rotation as indicated by ICP sustained higher than 20 torr.

The issue of turning rests on the judgment of the critical care nurse who must balance the effects of various ICP interventions with the effects of rotation. Sometimes actions to facilitate pulmonary function alleviate factors that increase ICP. Close monitoring and adjustment of rotational bed settings so that a patient rotates to two possible directions, from supine to the side of best ICP, can allow continued mobilization of the patient in the presence of precarious intracranial hypertension.

Figure 4. Photograph of the RotoRest® bed used at our institution. The bed is shown rotated about 45 degrees to the left.

Avoidance of Peripheral Nerve Injury

Patients with various degrees of brain injury may or may not be able to sense or respond to the numbness, tingling, or pain associated with the onset of a peripheral nerve injury. Much of the research on iatrogenic peripheral nerve injuries is based on monitoring of postoperative complications. The most common peripheral nerve injury is ulnar neuropathy, which is associated with positioning for surgery. As a preventive measure at our institution, neurosurgical patients undergo monitoring during surgery to identify slowing of nerve conduction that can signal the possible onset of neuropathy. When indicated by the EEG technician, patients are repositioned during surgery to prevent potential injury.

In other inpatient care settings where patients are not monitored by EEG, the preventive measures advocated by the Association of Operating Room Nurses and The American Society of Anesthesiologists can still be considered.[1] These measures include avoiding extended pressure on the ulnar (Fig. 5) and peroneal nerves (Fig. 6). Patients are positioned with the extremities supported on pillows while the extremes of flexion and extension are avoided. To prevent irritation of the ulnar nerve, pressure is avoided in the area of the elbow and ulnar groove. To prevent irritation of the peroneal nerve, pressure on the head of the fibula must be avoided. Therefore, pillows are placed to avoid pressure behind the knee. The variability of peripheral nerve anatomy among patients requires nurses to develop individualized positioning changes for each patient. The nurse can adjust positioning by recognizing subtle movements of a semicomatose patient such as the tendency to move an extremity frequently or to grimace when a limb is repositioned. Simply alternating the position of an upper extremity from semiprone to supine can alleviate pressure on a susceptible ulnar nerve.[3]

Cardiovascular Function, Joint Mobility, and Motor Function

The comatose condition of patients recovering from serious brain injury can lead to weakened cardiovascular function, joint immobility, and decreased motor function. Neurologic injury can lead to spasticity and abnormal muscle tone, and these symptoms can be exacerbated by immobility. Bedrest in healthy subjects is associated with an increased heart rate and decreased cardiac vagal tone. In a study of five young healthy males subjected to 21 days of bedrest, an increase in their maximal heart rate was associated with a reduction in their maximal uptake of oxygen. The authors related the findings to decreased stroke volume and cardiac output.[4] Periods of immobility can lead to stiffness; to shortening of tendons and musculature surrounding the joints, especially in the scapula, hip, and lower back;[2] and to bone loss that can be slow to reverse with therapy. Loss of joint mobility can impair rehabilitation. Patients with joint immobility must struggle to overcome both the immobility and paresis.

For these reasons, position changes have an important role in maintaining mobility for patients who will eventually regain motor function. Intermittently raising the head of the patient's bed to 45 to 90 degrees for short periods as soon as tolerated can help promote orthostatic responsiveness and combat some of the cardiovascular changes related to the supine position. Recognizing muscle hypertonicity and flaccidity before repositioning can help optimize intervention. Patients with hypertonic resistance to passive range of motion benefit from positioning that gently encourages relaxation of hypertonic muscles and increased range of motion of the affected joints. Some hemiplegic patients respond to positioning on the affected side so that the affected scapula is splinted in a position that decreases pain, allowing the surrounding muscles to relax. Positioning the upper extremities so that gravity can gradually open an affected elbow joint can enable a gentle increase in mobility of the upper extremity. Wrapping thin pillows to serve as splints around the elbow can help decrease hypertonicity and open the joint (Fig. 7).

Simple measures in turning brain-injured patients can prevent injury to joints and ensure smooth action of the muscles. When a patient is turned, first moving the patient's nose and chin into the direction of the turn often elicits a cooperative effort from the patient to reach into the turn (Fig. 8). By carefully placing a hand with fingers spread supportively behind the patient's scapula, the nurse can move the patient further into the sidelying position without injuring the scapula or shoulder joint.

Other measures are considered when a brain-injured patient is being repositioned. Periodically, patients slip down in bed and must be repositioned (Fig. 9). Incapacitated patients should be placed in a sitting position in bed or in a supportive chair at least twice a day. Consistently placing support under the affected

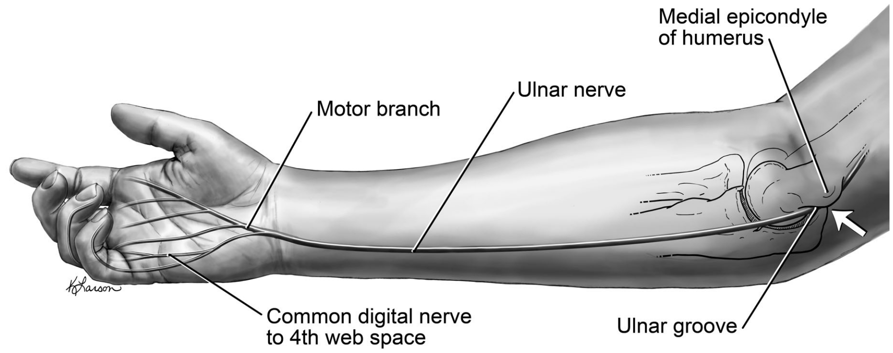

Figure 5. Illustration showing the course of the ulnar nerve and the site susceptible to pressure injury (*arrow*).

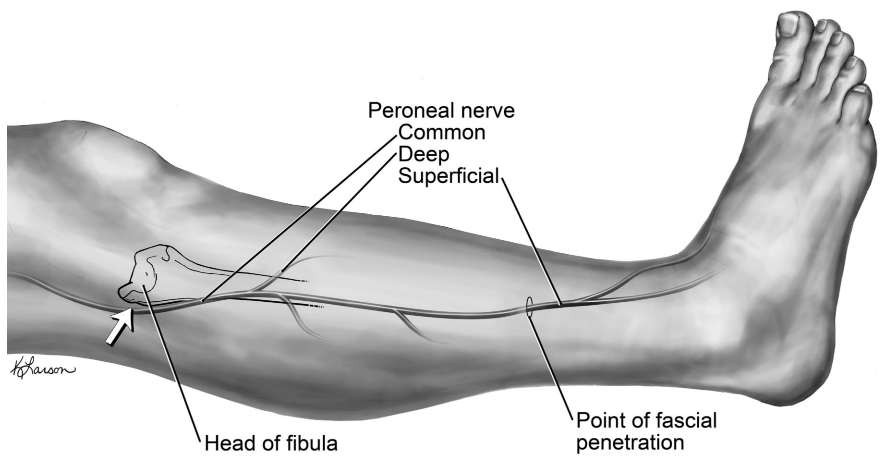

Figure 6. Illustration showing the course of the peroneal nerve in the lower leg and the site susceptible to pressure injury (*arrow*).

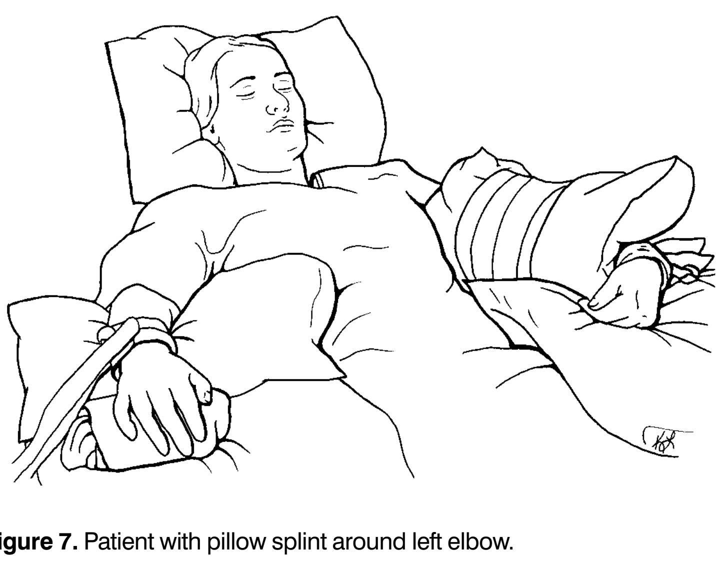

Figure 7. Patient with pillow splint around left elbow.

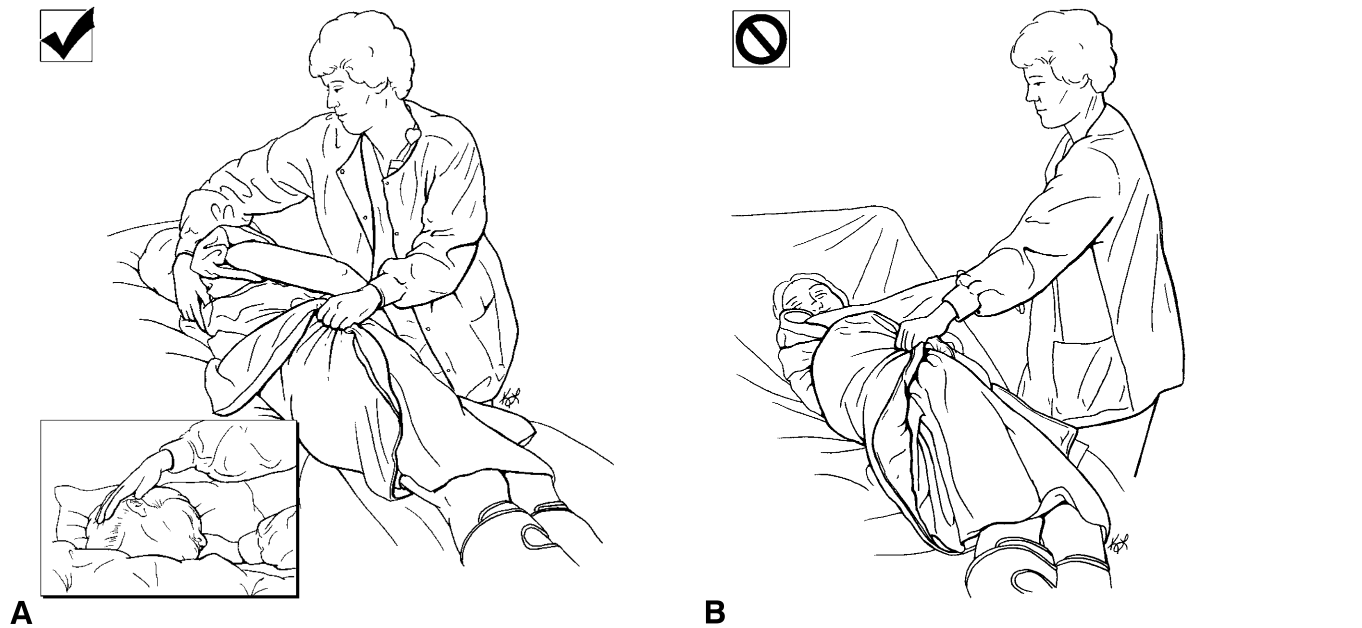

Figure 8. (*A*) Appropriate technique for turning incapacitated patients. The patient's head is first positioned in the direction of the turn (*inset*). (*B*) Inappropriate turning technique can lead to shoulder subluxation. When the head lags behind, the patient is more likely to resist the turn.

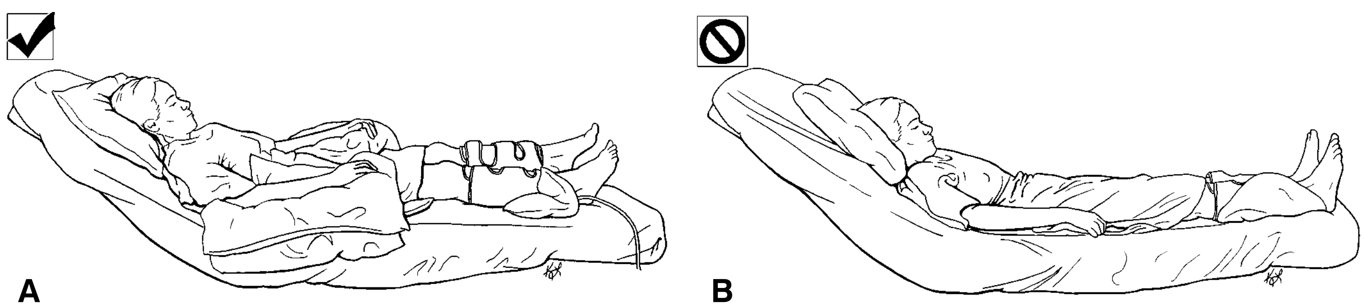

Figure 9. (*A*) When patient is positioned in bed appropriately, the hips are flexed to encourage normal expansion of the chest and abdomen. (*B*) When the patient's body slips toward the foot of the bed, flexion occurs higher than the hip and breathing is inhibited.

limb and behind both the affected and unaffected scapula can be enough to help the seated patient to maintain an aligned upright posture (Fig. 10). This position promotes chest expansion and decreases the likelihood of shoulder subluxation.

Early bilateral range of motion is another significant intervention provided by nurses and therapists. Range of motion performed bilaterally with the patient in a neutral supine position with the upper extremities moved in tandem may contribute to positive redevelopment of movement pathways (Fig. 11). Linking the reaching and rotating movements of the upper extremity during position changes strengthens core muscles and contributes to the balanced development of laterally weakened trunk muscles. This therapeutic maneuver can rehabilitate sitting balance and eventually can aid posture while standing. Proponents of this type of assisted activity stress the importance of avoiding overstimulation of compensatory unilateral hypertonic movements. Further increase in this kind of mobility is achieved by stimulation of normal body movements during activities of daily living and with activities that facilitate normal balance and muscle tone. Great patience and effort are needed to support patients in performing their own simple movements such as reaching toward a side rail or moving higher in bed.

Pulmonary Function

Managing patient position to optimize pulmonary function is a common challenge for neuroscience nurses, and current research supports the nurses' efforts in this area. Studies have shown that maintaining the head of the bed at a minimum of 30 degrees is independently associated with fewer occurrences of ventilator-acquired pneumonia.[9,12] Studies also support intermittently positioning patients in the prone position to promote

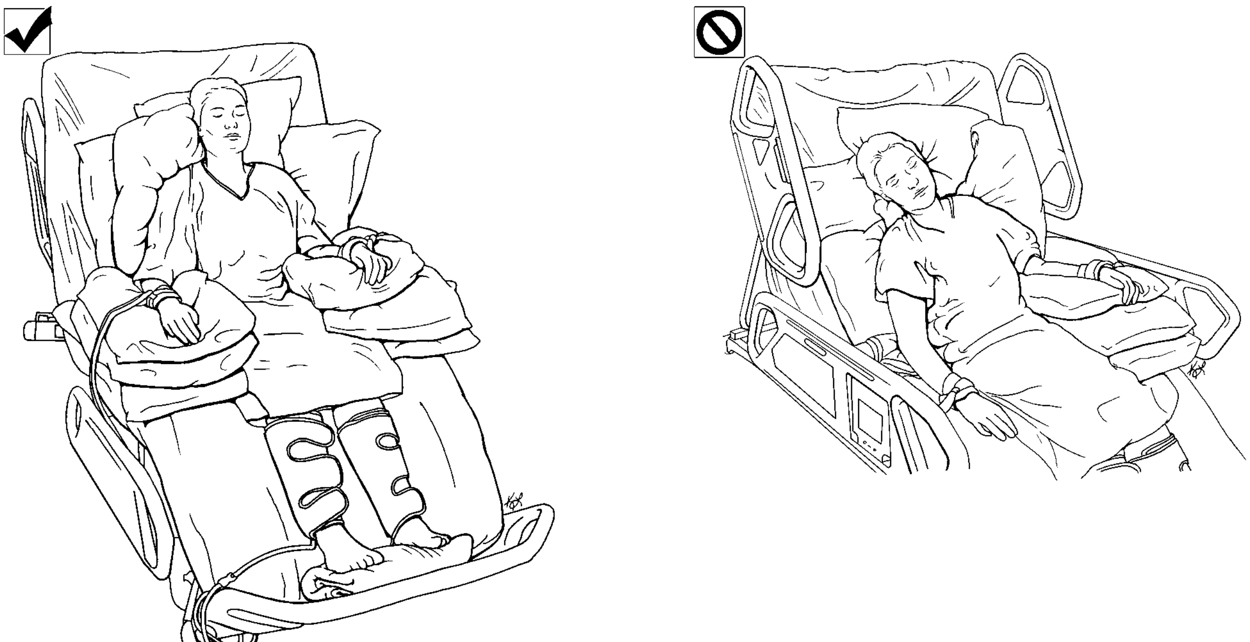

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Figure 10. (*A*) Appropriate positioning of a right hemiplegic patient. The right side of the head, both scapulae, and both arms are supported to maintain an aligned upright position. (*B*) When the patient is positioned inappropriately and the head and limb are unsupported, the patient lists to the right.

A B

Figure 11. Sequence of range-of-motion exercises. The nurse moves the patient's upper extremities in tandem.

recovery from severe respiratory complications.[5] The nurse must balance the positive effect of positioning against the patient's ability to tolerate the position. As stated earlier, small changes in position can have large effects on ICP. Anticipating how rotation will affect the position of invasive lines such as the endotracheal tube, ventilator tubing, feeding tube, and central line is important. Overstimulation of a cough or gag reflex by movement of the lines can cause precarious increases in lung pressure.

Adding a semiprone position to a turning regimen may be beneficial for patients unable to tolerate a fully prone position. The semi-prone position is similar to the side-lying position but includes more rotation (Fig. 12). The patient's elevated shoulder, arm, hip, and lower limb are rotated beyond midline and supported on pillows. This position is thought to help increase circulation within the lung fields and to allow some movement of static secretions, which, in turn, may open collapsed alveoli. Although the patient is not fully prone in this position, the patient's airway is still less visible to the nurse. Consequently, it is crucial to check the airway to make sure that it remains patent.

Figure 12. Appropriate placement of patient in the semi-prone position. Because the patient's airway is partially obscured, the nurse must be vigilant to ensure adequate respiration.

Cognition

Positioning procedures can have positive effects on the cognition of patients who are lethargic or even semicomatose. Such patients may have their eyes open only for brief periods. When reclined to 30 degrees or less, as is typical, their view is mostly of the ceiling and upper parts of walls (Fig. 13A). Caregivers are predominantly seen leaning over side rails straining to place themselves in the patient's line of vision. If the patient's response to position changes (i.e., ICP, cerebral perfusion pressure, systolic arterial blood pressure) begins to stabilize, repositioning the patient to an upright 45- to 90-degree angle provides a relatively normal view (Fig. 13B). Caregivers in the patient's line of vision no longer need to strain to see the patient. They can stand or sit comfortably and see the patient "eye to eye." This perspective may help patients to comprehend their environment, to see their family in ordinary stance, and thereby to improve their interaction with their caregivers.[7]

Figure 13. (*A*) Typical view from patient's perspective when the head of the bed is positioned at 30 degrees. (*B*) View from patient's perspective when positioned at a 90-degree angle.

Conclusion

Neuroscience nurses must balance the basic standards of nursing practice with evidence-based and aesthetic aspects of caregiving. The benefits of regular changes in a patient's position must be weighed against the risk of fluctuations in cerebral blood flow, cerebral perfusion pressure, ICP, and systolic arterial blood pressure and the myriad issues associated with immobility. When these complex issues are artfully balanced by neuroscience nurses, significant complications can be avoided and lifetime outcomes for patients can be improved.

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