

# Transarticular screw fixation of C1–2 for the treatment of arthropathy-associated occipital neuralgia

## Report of 2 cases

**PEYMAN PAKZABAN, M.D.**

*Houston MicroNeurosurgery, Houston, Texas*

Two patients with occipital neuralgia due to severe arthropathy of the C1–2 facet joint were treated using atlantoaxial fusion with transarticular screws without decompression of the C-2 nerve root. Both patients experienced immediate postoperative relief of occipital neuralgia. The resultant motion elimination at C1–2 eradicated not only the movement-evoked pain, but also the paroxysms of true occipital neuralgia occurring at rest. A possible pathophysiological explanation for this improvement is presented in the context of the ignition theory of neuralgic pain. This represents the first report of C1–2 transarticular screw fixation for the treatment of arthropathy-associated occipital neuralgia. (DOI: 10.3171/2010.10.SPINE09815)

**KEY WORDS** • **occipital neuralgia** • **C1–2 arthropathy** • **transarticular screw**

**S**ECONDARY occipital neuralgia associated with arthropathy of the C1–2 facet joint is a well-known phenomenon.<sup>7,20</sup> Transarticular screw fixation of C1–2 has been used for the treatment of severe neck pain (but not neuralgia) caused by C1–2 facet arthropathy in the absence of gross instability.<sup>8,10,11</sup> Here, I describe the first instances of C1–2 fusion with transarticular screws for the treatment of AAON. Importantly, the C-2 nerve root and ganglion were not decompressed in the 2 patients featured.

Occipital neuralgia due to reversible compressive syndromes of a vascular, neoplastic, and posttraumatic nature has been successfully treated with decompressive surgery.<sup>3,4,17,22</sup> It is reasonable to assume that the same pathophysiological mechanisms that mediate immediate postoperative improvement after microvascular decompression for trigeminal neuralgia also apply to decompressive surgery for occipital neuralgia. However, in the absence of decompression, improvement of occipital neuralgia with motion elimination alone requires further explanation. A possible pathophysiological mechanism for this improvement consistent with the ignition theory for trigeminal neuralgia is provided below.<sup>5,16</sup>

*Abbreviation used in this paper:* AAON = arthropathy-associated occipital neuralgia.

## Case Reports

### *Case 1*

*History and Examination.* This 55-year-old woman with rheumatoid arthritis presented with an 18-month history of severe, episodic electrical-type pain radiating from the left side of the neck to the left occipital and retroauricular regions of the scalp. This paroxysmal pain was superimposed on a dull constant left-sided neck pain. The paroxysmal pain was reproducibly provoked by neck movements, particularly neck extension. Just prior to surgery, spontaneous paroxysms would occur after 20 minutes of sitting or standing, even without gross neck movements, requiring the patient to lie on her back to alleviate the pain. A soft cervical collar reduced the frequency of the attacks. On examination, exquisite tenderness and a positive Tinel sign were noted in the region of the left C1–2 facet joint in an otherwise neurologically intact patient.

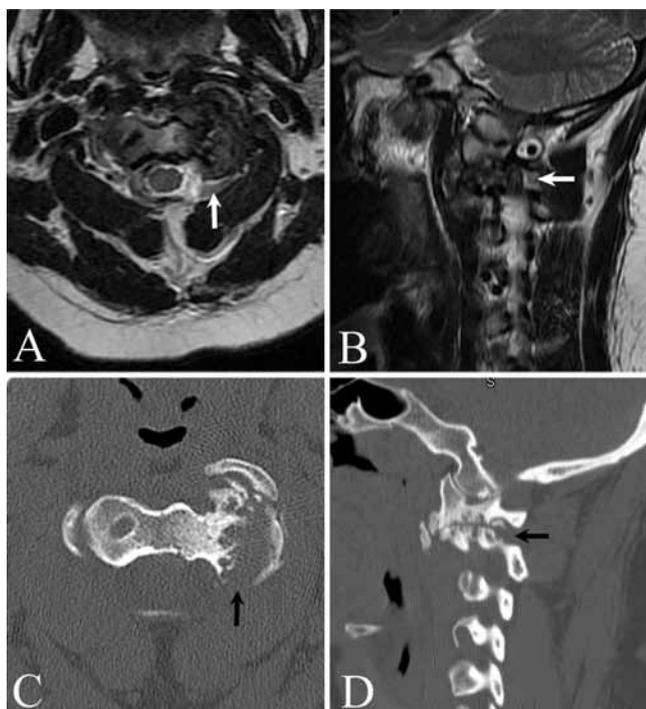
Her condition had not responded to treatment with naproxen, ibuprofen, oral corticosteroids, hydrocodone, oxycodone, gabapentin, and carbamazepine. She had no pain relief after an occipital nerve block and attempted alcohol ablation of the left occipital nerve. She had received 3 injections of bupivacaine and methylprednisolone at the

left C1–2 facet joint, producing transient improvement of the neck pain but not the occipital neuralgia.

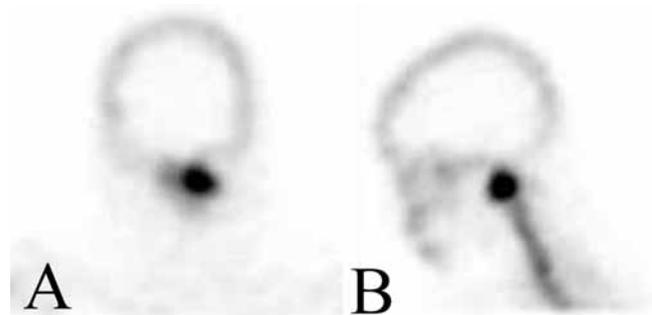
Magnetic resonance imaging (Fig. 1A and B) revealed marked unilateral hypertrophy of the left C1–2 facet joint, producing posterior displacement of the C-2 nerve root. Computed tomography scanning (Fig. 1C and D) revealed extensive destruction of the left C1–2 articulation with partial osteolysis of the C-1 and C-2 lateral masses, accompanied by marked bony hypertrophy and osteophyte formation in the C-2 nerve root recess. Bone scanning (Fig. 2) revealed intense focal radionuclide uptake in the region of the left C1–2 facet joint. Flexion and extension cervical radiographs excluded gross atlantoaxial instability.

**Operation.** After bilateral posterior exposure of the C1–2 region, the left C-2 nerve root and ganglion were found to be markedly stretched over a broad-based osteophyte arising from the C1–2 facet joint, which did not lend itself to resection. A biopsy sample of the soft tissue within the left C1–2 articulation was taken and yielded chronic inflammatory tissue on pathological examination. A transarticular screw was placed on the right side. A screw could not be placed on the left side because of the destruction of the left C-2 pars and substantial portions of the C-1 and C-2 lateral masses. A C1–2 fusion with Sonntag-style cable fixation was performed.<sup>15,19</sup>

**Postoperative Course.** The neuralgic pain paroxysms, both spontaneous and motion-induced, resolved



**FIG. 1.** Case 1. Axial (A) and sagittal (B) T2-weighted MR images depicting marked unilateral hypertrophy of the C1–2 facet joint and compression and posterior displacement of the C-2 nerve root (white arrows). Axial (C) and sagittal (D) CT reconstructions depicting the coexistence of destructive and hypertrophic changes in the C1–2 facet joint. Black arrows represent the expected location of the C-2 nerve root.



**FIG. 2.** Case 1. Coronal (A) and sagittal (B) bone scans demonstrating intense focal radionuclide uptake in the region of the left C1–2 articulation.

immediately after the operation and have not recurred after 24 months of follow-up. The patient's neck pain gradually improved and resolved within 6 weeks. All narcotics and anticonvulsants were tapered and discontinued. She remained on infliximab and naproxen for maintenance treatment of rheumatoid arthritis. Solid fusion was confirmed on CT at 6 months postoperatively (Fig. 3).

#### Case 2

**Presentation.** This 57-year-old woman with osteoarthritis presented with a 12-month history of severe, paroxysmal stabbing pain radiating from the left suboccipital region to the left side of the vertex. She had suffered from chronic neck pain for 5 years prior to the onset of occipital neuralgia. The neuralgic pain occurred at rest and was further provoked by neck movements. She had remained in bed for 3 weeks prior to surgery because of the frequency and severity of the pain paroxysms. On examination, she held her neck rigid and resisted any movement. A positive Tinel sign was elicited at the occipitocervical junction on the left side. She had no neurological deficits.

Prior to the onset of occipital neuralgia, the patient had undergone several courses of physical therapy and cervical epidural steroid injections over a 5-year period for the treatment of neck pain. She had been taking propoxyphene and various nonsteroidal antiinflammatory drugs on an intermittent basis. After the onset of occipital neuralgia, she underwent treatment with pregabalin, oxcarbazepine, and Carbatrol without benefit. Her narcotic requirements escalated to large doses of hydromorphone and fentanyl transdermal patches. She did not respond to an occipital nerve block or a trial of percutaneous peripheral nerve stimulation of the left occipital nerve, disqualifying her for internalization of the stimulator.

Magnetic resonance and CT studies (Fig. 4A and B) revealed exuberant degenerative hypertrophy of the left C1–2 facet joint and the presence of a broad-based osteophyte in the C-2 nerve root recess that compressed and displaced that root. Correspondingly, a bone scan (Fig. 4C) revealed intense radionuclide uptake in this region. Dynamic radiographs excluded gross C1–2 instability.

**Operation.** The left C-2 nerve root and ganglion were markedly stretched over a large broad-based osteophyte arising from the C1–2 facet joint. Adequate decompression of the C-2 nerve root was not believed to be feasible



**FIG. 3.** Case 1. Images obtained 6 months after surgery. Sagittal CT reconstructions (A–C) across the left facet joint, midline, and right facet joint, respectively, demonstrating fusion across the uninstrumented arthritic left facet joint, fusion between the odontoid and anterior C-1 arch, and fusion of the posterior bone graft. Three-dimensional CT reconstruction (D) demonstrating an anterior view of the fused arthritic joint on the left side. Lateral plain radiograph (E) showing fusion constructs. Black arrows correspond to fusion interfaces.

and thus was not attempted. Bilateral atlantoaxial transarticular screw fixation supplemented by a modified Sonntag fusion was performed (Fig. 5).

**Postoperative Course.** The patient's occipital neuralgia resolved immediately and completely after the operation. Anticonvulsants were rapidly tapered and discontinued. Narcotics were tapered back to propoxyphene within 3 weeks and were discontinued completely after 8 weeks. Solid fusion was confirmed on CT at 3 months (Fig. 6). She was pain free at the last follow-up, 22 months after surgery.

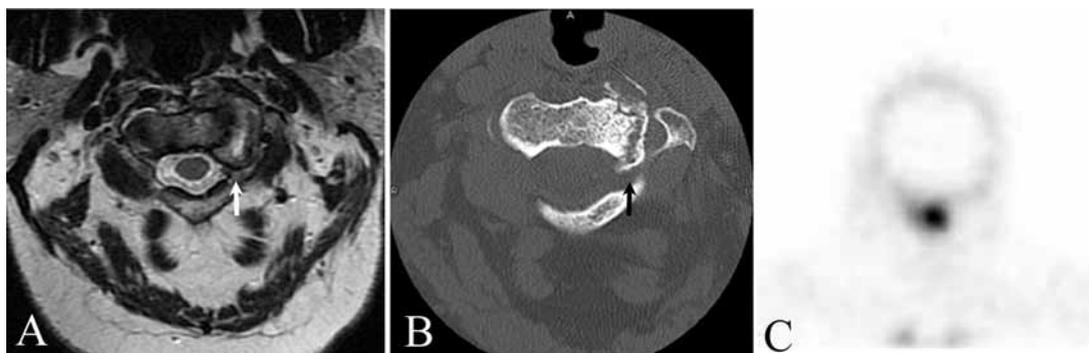
### Discussion

These 2 patients presented with refractory occipital neuralgia and clear evidence of ipsilateral C1–2 arthropathy, satisfying the definition of AAON.<sup>7</sup> They experienced true occipital neuralgia (unpredictable bursts of intense paroxysmal pain occurring at rest) and movement-evoked C-2 radicular pain, and both resolved immediately after

C1–2 stabilization, as did the chronic arthritic neck pain. Importantly, decompression or section of the C-2 nerve root was not performed in either case. These findings provide not only an alternative treatment strategy for AAON, but also an insight into the pathophysiology of this disorder.

### Implications for Treatment

When Ehni and Benner<sup>7</sup> first described occipital neuralgia associated with C1–2 arthrosis in 1984, they recommended C-2 dorsal rhizotomy for treatment, recognizing the infeasibility of a C-2 nerve root decompression. One of the patients in that report underwent both a C-2 rhizotomy and a C1–2 fusion, making it impossible to determine the relative contribution of each therapeutic strategy to the resolution of pain. A C-2 rhizotomy and ganglionectomy have shown variable success in the long-term control of idiopathic occipital neuralgia,<sup>1,12,18,21</sup> do not address the primary pathology in AAON and cannot be expected to treat the nonneuralgic arthritic neck pain in AAON.



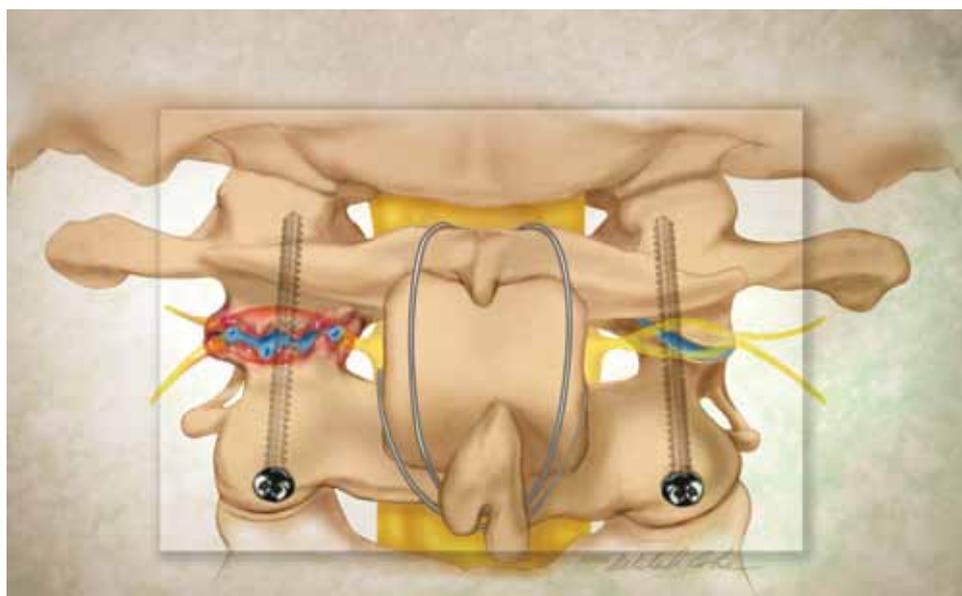
**FIG. 4.** Case 2. **A:** Axial T2-weighted MR image depicting unilateral hypertrophy of the C1–2 facet joint and compression of the C-2 nerve root (*white arrow*). **B:** Computed tomography scan demonstrating osteophyte formation within the C-2 nerve root recess. *Black arrow* depicts the expected location of the C-2 nerve root. **C:** Coronal bone scan demonstrating focal radionuclide uptake in the expected location of the left C1–2 facet joint.

The novelty of the present report rests neither in using C1–2 instrumented fusion for the treatment of painful facet arthropathy, nor in reporting successful resolution of cervical radicular pain by the immobilization of a cervical motion segment. Rather, this is the first report of the successful treatment of true occipital neuralgia via C1–2 fusion. To fully appreciate the implications of these findings, it is important to make a distinction between neuralgia and compressive radicular pain and, in particular, between occipital neuralgia and C-2 radiculopathy. In neuralgia, the characteristic short-lived explosions of intense pain are caused by a positive feedback loop that results in the rapid and exponential recruitment of aberrant axonal afterdischarges.<sup>5</sup> In the instances that decompression successfully treats neuralgic pain, it may do so by eliminating the ignition stimulus, as described below. In contrast, in compressive radiculopathy, the more constant nature of the pain, the predictable exacerbation of pain by maneuvers that further distort the nerve root, and the frequent resolution of such pain after decompression

reflect a simpler, more direct relationship between axonal distortion and pain, which in turn explains the more predictable response of this type of pain to decompression.

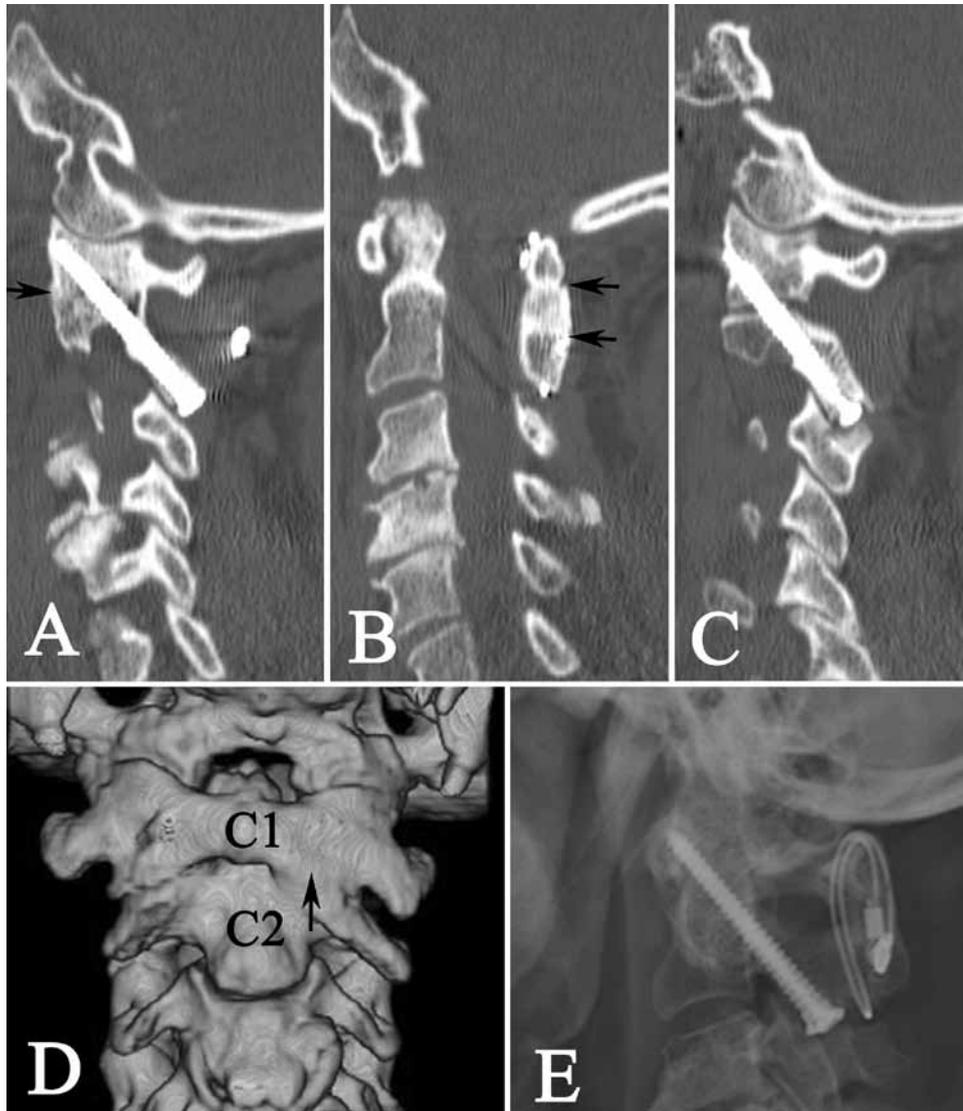
An exhaustive literature search revealed only a single published report of atlantoaxial fusion for presumed occipital neuralgia, which on closer inspection turned out to be a C-2 radicular pain syndrome.<sup>13</sup> Specifically, this brief report describes the occurrence of C-2 radicular pain in laborers who carried heavy loads on their heads. The resolution of this load-dependent radicular pain 3 months after C1–2 fusion reflects shielding of the C-2 nerve root by the fusion construct against axial compression rather than the treatment of true occipital neuralgia. In contrast, the present study represents the first report of the immediate resolution of true occipital neuralgia after C1–2 instrumented fusion without decompression.

It is important not to generalize these findings to other types of occipital neuralgia. Clearly, C1–2 fusion is not a treatment for idiopathic occipital neuralgia, nor should it be the first line of treatment for patients with AAON.



**FIG. 5.** Artist's representation of C1–2 fusion with transarticular screw fixation for the treatment of AAON. Notice that the symptomatic left C-2 nerve root and ganglion (*in red*) have not been decompressed. Reprinted with permission from the artist, Delilah R. Cohn. Source: <http://www.medillustrationstudio.com>.

## Fusion of C1–2 for occipital neuralgia



**FIG. 6.** Case 2. Imaging studies obtained 3 months after surgery. **A–C:** Computed tomography sagittal reconstructions across the left facet joint, midline, and right facet joint, respectively, demonstrating fusion across the arthritic left facet joint and fusion of the posterior bone graft. **D:** Three-dimensional CT reconstruction demonstrating an anterior view of the fused arthritic joint on the left side. **E:** Lateral plain radiograph showing fusion construct. *Black arrows* correspond to fusion interfaces.

Most patients with AAON are successfully treated using conservative means, and many do not even get referred for neurosurgical evaluation. A C1–2 fusion should be reserved as a last resort for patients with clearly identifiable C1–2 facet arthropathy and severe concordant occipital neuralgia whose condition has failed all nonsurgical (and possibly other surgical) measures, as in the 2 cases presented here. Even in such a highly selected group, this treatment strategy represents a novel approach that is far from accepted.

### *Pathophysiological Implications*

An interesting and unexpected finding in these patients was the immediacy with which occipital neuralgia resolved after joint stabilization. After all, no decompression was performed, and the C-2 nerve root and ganglion could not have healed from any putative chronic injury

within this time frame. Why would motion elimination address not only motion-evoked pain, but also neuralgia at rest?

A possible hypothesis is proposed by analogy with trigeminal neuralgia. In trigeminal neuralgia, demyelination of sensory axons compressed by arterial loops has been well documented.<sup>5</sup> The ignition hypothesis explains how demyelination can result in paroxysmal pain and why such pain is relieved immediately after microvascular decompression.<sup>16</sup> Injured neurons are susceptible to spontaneous or stimulus-induced afterdischarge. Demyelination results in axon-to-axon cross-excitation.<sup>2</sup> Ephaptic cross-talk and crossed afterdischarges result in the synchronization of afterdischarges in a large group of neurons.<sup>6</sup> Recruitments of increasingly large groups of axons in this process produce a chain reaction that terminates in an explosion of pain.<sup>5</sup> Importantly, although chronic compress-

sion of the nerve root is responsible for demyelination, it is the pulsation of the artery against the demyelinated nerve root that is thought to produce the ignition stimulus for the pain paroxysms. The fact that not every pulsation produces a paroxysm has to do with that time constant for the refractory period after massive axonal depolarization.<sup>5</sup> It is, therefore, the elimination of pulsatility and not decompression that accounts for immediate pain relief after microvascular decompression (long before any healing of the injured axons can occur).<sup>9,14</sup> Importantly, rare instances of occipital neuralgia due to vascular compression have been successfully treated with microvascular decompression.<sup>17,22</sup> The immediate relief of paroxysmal pain in these instances suggests that the C-2 nerve root may respond in a similar fashion as the trigeminal nerve root to the elimination of pulsatility.

The same mechanism can be used to explain the immediate resolution of paroxysms of occipital neuralgia in the 2 patients in the present report, even though the C-2 nerve root and ganglion were not decompressed. Specifically, chronic compression of the C-2 nerve root and ganglion by the hypertrophic facet joint can be expected to produce axonal injury and demyelination, whereas movement (including micromotion) of the diseased facet joint against the injured nerve root may provide the ignition stimulus. The exclusion of such movements by C1–2 transarticular screw fixation may eliminate the ignition stimulus, thereby relieving the paroxysms of neuralgic pain.

It is unclear whether similar pathophysiological mechanisms are operative in idiopathic occipital neuralgia.

#### Diagnostic Implications

We found that the combination of CT and MR imaging not only identified isolated unilateral C1–2 facet arthropathy, but also definitively demonstrated compression of the C-2 nerve root in these patients with AAON. Although dynamic radiographs excluded gross instability in the patients, it appears reasonable to obtain these images in all patients suspected of suffering from AAON. The dramatic focal radionuclide uptake by the offending joint in the 2 patients featured suggests that a bone scan may serve as a useful screening test for AAON. The excellent response of AAON to C1–2 fusion in this study further reinforces the need for careful radiographic evaluation of the upper cervical spine in all patients with refractory occipital neuralgia, lest they suffer from treatable AAON.

#### Disclosure

The author serves as a consultant for Aesculap Implant Systems, Inc., and Synthes USA Products, LLC.

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Address correspondence to: Peyman Pakzaban, M.D., 3801 Vista, Suite 440, Pasadena, Texas 77504. email: pakzaban@swbell.net.