Obstetric Forum

Epidural analgesia for labour in a parturient with neurofibromatosis

Mokhtar Dounas MD, Frédéric J. Mercier MD, Cécile Lhuissier MD, Dan Benhamou MD

The first report of epidural analgesia for labour in a 26-yr-old woman with von Recklinghausen's neurofibromatosis is described. Epidural anaesthesia is often considered as contraindicated because neurofibromas may involve spinal cord and nerve roots. However, general anaesthesia was considered at high risk for this parturient on the basis of her previous medical and surgical history and of physical findings. The present observation suggests that epidural analgesia may be used in such circumstances provided that spinal cord neurofibromas have been ruled out by clinical and CT scan (or magnetic resonance imaging) examination.

Les auteurs rapportent le premier cas d'analgésie péridurale pour l'accouchement chez une parturiente de 26 ans atteinte de neurofibromatose de Von Recklinghausen. L'analgésie péridurale est déconseillée voire contre-indiquée pour de nombreux auteurs en raison de la possible localisation radiculaire des neurofibromes. Cependant cette patiente a un lourd passé médico-chirurgical rendant la pratique de l'anesthésie générale très risquée. Cette observation montre que l'analgésie péridurale peut être utilisée dans cette situation, à condition d'avoir éliminé par un examen clinique et radiologique (scanner ou IRM) la localisation d'un neurofibrome sur les racines lombaires correspondant à l'espace de ponction.

Key words
ANAESTHESIA: obstetrical;
ANAESTHETIC TECHNIQUES: epidural;
COMPLICATIONS: neurofibromatosis.

From the Département d'Anesthésie-Réanimation, Hôpital Antoine Béclère, 157 rue de la Porte de Trivaux, 92141 - Clamart Cedex - France.

Address correspondence to: Dr. D. Benhamou, Département d'Anesthésie-Réanimation, Hôpital Antoine Béclère, 157 rue de la Porte de Trivaux, 92141 - Clamart Cedex - France.

Accepted for publication 1st January, 1995.

Von Recklinghausen's neurofibromatosis is an inherited autosomal dominant disease with a reported incidence of about one in 3000 births. This disease is always progressive but with a markedly variable expressivity. It is characterised by abnormal cutaneous pigmentation ("café au lait" spots) and numerous neurofibromas of peripheral nerves.[1] Neurofibromatosis manifestations may not only worsen during pregnancy[2-5] but also may have deleterious effects on pregnancy.[1,5]

Epidural analgesia is the most effective method to provide pain relief for labour.[6] Although several authors consider that epidural analgesia may be at risk or even contraindicated in the presence of neurofibromatosis,[7,8] we are unaware of specific references about anaesthetic management of these parturients. Thus, this is the first case report describing the successful use of epidural analgesia for labour in a parturient with neurofibromatosis.

Case report

A 26-yr-old primigravida was scheduled for an anaesthetic visit at 31 wk gestation. Her past medical history included the diagnosis of Von Recklinghausen's neurofibromatosis during childhood revealed by a left supraclavicular compressive tumour. The patient underwent surgery but tumour removal was incomplete because of cervical vertebral adhesion. A second operation was undertaken four years later for recurrence. Surgery was technically difficult and also incomplete; the collar-bone and the first left rib were excised and several nerve roots and the left phrenic nerve were cut. Four other operations were undertaken later for recurrences. An evaluation, ten years ago, showed left cervical scoliosis of 20°, right thoracic scoliosis of 30°, left phrenic nerve paralysis and a restrictive syndrome (vital capacity = 1600 ml – predicted 3400 ml, residual volume = 400 ml – predicted 800 ml, and FEV_1/VC = 70%). Pregnancy continued normally until 30 wk gestation. At this time, preterm labour occurred but was easily controlled with bed rest and progesterone administration. Pelvimetry showed an adequate pelvis for vaginal delivery. Airway evaluation revealed limited neck extension and a partial view of the

oropharynx (Mallampati class 2).[9] Deviation of the trachea to the right was noted on the chest *x*-ray. As previous surgery had been incomplete, a thoracic CT scan (General Electric 9800, Milwaukee, USA) with radiation abdominal protection was performed to evaluate the spread of the remaining tumour in the thorax. It showed a 10-cm diameter mass on the left paratracheal mediastinum with a consequently compressed lung parenchyma. A lumbar CT scan with a limited number of views was also performed to visualise the region that would be involved by epidural puncture. It showed an abnormal image facing the second lumbar vertebrae, which evoked a subarachnoid neurofibroma. However, structures below L_2 were normal.

The patient was admitted to hospital for labour at 36 wk gestation. Preoperative laboratory tests (Hb, Hct, WBC, platelet count, fibrinogen, prothrombin time) were normal. Heart rate and noninvasive arterial blood pressure were monitored and prehydration was performed with 500 ml crystalloid. After skin anaesthesia with 2 ml lidocaine 2%, a midline puncture was made using a 18-gauge Tuohy needle. The L_4-L_5 interspace was chosen because lumbar CT scan showed that this interspace was free of neurofibroma. The puncture was difficult because of the marked scoliosis and several attempts were necessary to succeed. The epidural space was identified by loss of resistance to saline 4 cm from the skin. The catheter was easily introduced into the epidural space, without paraesthesia. Following negative aspiration for both blood or cerebrospinal fluid, 15 ml bupivacaine 0.125% was injected in 5-ml increments over six minutes. Then, a continuous infusion with the same concentration was started at a rate of 15 $ml \cdot hr^{-1}$. After a nine hour labour, a healthy neonate was delivered (Apgar scores 10 and 10), by low pelvis instrumental delivery. The postdelivery course was uneventful and the patient was discharged five days later.

Discussion

Neurofibromatosis may affect multiple organs,[1] and thus may have several implications for the obstetrical anaesthetist. The prenatal anaesthesia visit should focus on the airway. Difficult tracheal intubation may be expected because of stiffness of the cervical spine, deformity of the facial bones (with asymmetry, hypertrophy or atrophy), macroglossia, and tumour involving the tongue or the larynx.[10-12] Moreover, in these latter cases, manual ventilation after induction of general anaesthesia may be impossible because of oropharyngeal obstruction, leading to emergency cricothyroidotomy.[10] It must always be kept in mind that phaeochromocytoma may be associated in one percent of cases.[1] Indeed, during pregnancy, phaeochromocytoma may mimic pregnancy-induced hypertension and therefore the correct diagnosis is often made in the operating room or during post-mortem examination.[13] Vascular abnormalities are also an important feature of the disease. They may involve vessels of any calibre and may be either aneurysms or stenosis.[8] Hypertension is the most common consequence.[5] However, several deaths due to aneurysm rupture have been reported.[2-4] Therefore, some authors have suggested that hypertension should always be investigated thoroughly and computerised angiography done to eliminate aneurysms.[8] Therefore, if general anaesthesia is required, care must be taken to prevent any increase in arterial blood pressure particularly during laryngoscopy and intubation. Neurological complications should also be systematically searched far as Rubenstein *et al.*[14] found an incidence of 35.2% in a population of 250 patients with neurofibromatosis. Among these neurological complications, 7.2% were brain tumours and 5.6% were symptomatic spinal cord or nerve root neurofibromas. Symptomatic orthopaedic problems (essentially scoliosis) are also frequently encountered (6.8%).[14]

Among the anaesthetic agents commonly used, only muscle relaxants may be of concern. Several authors have suggested a prolonged duration of action of both depolarising and nondepolarising muscle relaxants.[15,16] However, by recording of the twitch response to ulnar nerve stimulation, Baraka documented a nearly complete resistance to 100 mg succinylcholine (and the lack of fasciculation), followed by a prolonged hypersensitivity to 30 mg of d-tubocurarine. The author pointed out that this kind of paradoxical response was rather similar to the response which had already been noted in patients with myasthenia gravis and, therefore, might be due to presynaptic defect and/or postsynaptic alteration of the endplate chemoreceptors' response.[17] Thus, neuromuscular monitoring should always be instituted when general anaesthesia with muscle relaxants is required for patients with neurofibromatosis. Instrumental delivery is usual to avoid expulsive efforts that may lead to rupture of an eventual aneurysm. Anaesthesia is therefore often required during delivery in patients with neurofibromatosis.

For this patient, there were several important arguments to avoid general anaesthesia. The stiffness of the cervical spine and the pronounced tracheal deviation suggested an increased risk of difficult intubation. If this had occurred, life-threatening hypoxaemia and fetal distress might have rapidly ensued because of the restrictive pulmonary syndrome associated with phrenic nerve paralysis. The efficacy of succinycholine to facilitate intubation for rapid-sequence induction might have been difficult to predict. There was no clinical argument suggesting a risk of complete obstruction of the oropharynx. However, when mask ventilation is needed to avoid

hypoxaemia after failed intubation, there is an important risk of pulmonary aspiration of gastric contents. Therefore, epidural analgesia was preferred and performed at L_4-L_5 interspace after a neurofibroma at this lumbar level was eliminated by a CT scan. Beside the technical difficulties often encountered in the presence of scoliosis, no complications occurred.

Our radiologist estimated that the dose delivered to the fetus during the seventeen abdominal views was about 10 mGy. This irradiation is the upper limit accepted before 10 wk of gestation but is far less than the 50 mGy threshold settled at the end of pregnancy.[18,19] Magnetic resonance imaging (MRI) was not used because of the presence of metal clips on large vessels into the thorax. However, critical analysis suggests that MRI should have probably been preferred since MRI may in fact have less deleterious effects than CT scan imaging provided contraindications to MRI are respected.

In conclusion, this observation highlights the use of epidural analgesia for labour in patients with neurofibromatosis. Instrumental delivery is frequent and therefore anaesthesia is often required. However, general anaesthesia may be particularly hazardous because of the high risk of difficult intubation and life-threatening hypoxaemia. Therefore, epidural anaesthesia may be proposed alternatively, provided that the presence of a neurofibroma at the puncture site has been ruled out by a careful clinical and radiological (CT scan or magnetic resonance imaging) examination.

COMMENTARY

Chantal Crochetière MD
Assistant Professor, Département d'Anesthésie, Hôpital Sainte-Justine, Montreal, Quebec

In 1994, do contraindications to regional anaesthesia in obstetrics still exist? We make subtle distinctions between hypovolaemia and compensated hypovolaemia, infection and localized infection, neurological problems and stable neurological deficit, biological coagulopathy and "clinical" coagulopathy, etc. Previous relative contraindications such as herniated disc in labour are becoming indications!

Epidurals are very safe in the obstetrical population and offer many advantages.[20] Even if an anaesthetist does not see a serious complication in one's own lifetime, that does not mean that it can never happen. Problems occur, and when they do they involve a young woman with a family to raise.

In this case report Dounas *et al.* there are many interesting points that are worth discussing.

Antenatal visits

We cannot stress enough the importance of antepartum evaluation of women with medical problems. Unfortunately, our colleagues in obstetrics are not always aware of potential problems related to anaesthesia. Clarifying a situation and obtaining informed consent during active labour at 3 a.m. can be distressing for both parties and the final decision is not always in favour of the patient. In a recent editorial, Smith talked about "preconception clinics" where women with chronic diseases could be educated about the impact a pregnancy may impose on their health and the resulting changes in the management of their disorder.[21] This would be the ideal world!

Radiologic investigation in pregnancy

Most radiologists in North America are reluctant to do a scan or a magnetic resonance imaging during pregnancy unless there is a strong indication, such as a progressive neurological deficit. Even with the benefit of a thorough pre-pregnancy investigation one cannot assume that the condition will be the same at the end of pregnancy. In this case, the patient did not have any new neurological symptoms and in our hospital she would have had a lumbar scan.

Difficult airway and regional anaesthesia

I feel that a proved or suspected difficult airway is an indication for regional anaesthesia. Even though difficult intubation is an important problem in obstetrics (main cause of "anaesthetic death" in this field),[22] one should keep in mind that lumbar spine involvement, as in this patient, could have prevented regional anaesthesia. Therefore, one must be ready to manage a difficult intubation. Mallampati classification should not be considered alone. The most important additional factors, especially with a Mallampati Class 2, are: short neck, receding mandible, and protruding maxillary incisors.[23] In this case, the patient had at least six previous operations. Did she have a previous airway problem?

Von Recklinghausen disease

Most of von Recklinghausen patients have mild disease that does not preclude any type of anaesthesia. It is rare to see a patient as seriously compromised as this lady at such a young age. Apart from the fact that she might have an abnormal response to muscle relaxants (this could be known from previous anaesthesia) and has a restrictive syndrome which could require postoperative ventilation, the main question is, "Do you offer an epidural to a patient with spinal neurofibroma and a ten percent chance of an intracranial tumour?" Careful examination by a neurologist should eliminate most important lesions but a spinal lesion can be asymptomatic

(compensated lesion). Paraplegia or even quadriplegia can develop after an accidental dural puncture (more frequent with abnormal anatomy), secondary to rapid decompression. If you decide to proceed, the patient should be evaluated regularly and should receive a low-dose continuous epidural infusion to prevent motor blockade.

Informed consent

There are three legal elements to an informed consent.[24]

1 adequate information given to the patient;
2 adequate understanding of the information by the patient;
3 voluntary decision by the patient.

In this kind of case, I think a physician cannot provide too much information and should carefully record the discussion in the chart. As well, opinions should be sought from experts in related fields; in this situation, a neurologist and neurosurgeon.

In difficult cases such as this, one has to weigh the benefits against the risks. This patient had a history of premature labour and evidence of an adequate pelvis, but one can never predict the outcome of labour. Initially her pain could have been managed conservatively, i.e., opioids or nitrous oxide/oxygen and this may have allowed an assessment of the pattern of labour and its possible result (vaginal delivery or Caesarean section). Unfortunately, I would not have had the benefit of a lumbar scan and, based on her past history, I would have been reluctant to perform an epidural due to a lumbar neurofibroma. Although neurofibromas are usually lateral, they can be large and asymptomatic. My major concern is not that of puncturing a neurofibroma, but rather the risk of accidental dural puncture with its potentially disasterous consequences. Again, I would be prepared for an awake fibreoptic intubation at any time should an emergency arise.

COMMENTARY

Holly Muir MD FRCPC
Staff Anaesthetist, Grace Maternity Hospital, Halifax, Nova Scotia

I found this case report of management of analgesia for labour in a 26-yr-old woman with von Recklinghausen's neurofibromatosis very thought-provoking. The authors present most of the features of the disease which make it hazardous in pregnancy. However, I feel that the unpredictable course of the disease in pregnancy should be emphasized; in particular, the tendency for rapid growth of neurofibromata and the risks of developing intracranial hypertension, haemorrhage and sarcomatous degeneration. This necessitates careful re-evaluation of a patient who presents for anaesthesia and should lead one to always question the validity of CT or MRI scans done on these patients before or early in pregnancy.

The choice of anaesthetic technique should be made on an individual basis between anaesthetist and patient, with all the known relevant information available. For many conditions, especially when neurological disease is involved, regional anaesthesia has been considered to be contraindicated simply because little is known about the disease state or the effects of regional anaesthesia on it. I am grateful to the authors for sharing their experience with us. In their report, they shared their decision-making processes, weighing the risks of general anaesthesia with a potentially difficult airway versus the risks and benefits of regional anaesthesia.

The provision of regional anaesthesia for relief of labour pain and instrumental delivery was beneficial to this particular patient. However, it could have come at some cost. There was still the risk that her disease could have progressed beyond that documented on a remote CT scan. Was an increase in intracranial pressure (ICP) adequately ruled out and the consequence of dural tap and persistant leak on a large intracranial lesion or aneurym discussed? Other questions that remain unanswered are: (1) How far would one insert the epidural catheter in the patient and should one be concerned about catheter migration and encroachment on diseased areas? (2) Could this patient's labour pain have been managed with opioid infusion, and delivery achieved with a pudendal or saddle block? (3) If Caesarean section had become necessary, would this patient have tolerated further respiratory compromise with an even greater restrictive syndrome imposed by respiratory muscle paralysis secondary to a regional block?

The above are worthwhile considerations in this patient. For labour, I would start with the technique with the least risk, i.e., good prenatal instruction in relaxation techniques, possibly transcutaneous electrical nerve stimulation (TENS) for early labour, and opioids and N_2O/O_2 for advanced labour, with the use of pudendal or saddle block for instrumental delivery. If these techniques failed and after detailed pre-labour discussion and consent, I would use epidural anaesthesia. I would feel very nervous about using a regional block for Caesarean section with her pre-existing respiratory compromise and would be happier with a plan for awake fibreoptic intubation and controlled ventilation with facilities for postoperative observation. The risks that this patient poses and the management plan should be discussed with the obstetrician, so that surgical intervention (Caesarean sec-

tion) could be performed in a controlled fashion, rather than waiting until the situation becomes extreme.

I would like to emphasize that my hesitancy to use regional anaesthesia for Caesarean section arises from her severe restrictive lung disease and not from her neurological condition. The majority of patients with von Recklinghausen's neurofibromatosis have very benign disease. In those who have spinal lesions, if recent radiological examination can rule out neurofibromas at the planned insertion site, regional anaesthesia can be safely administered and in the majority of obstetric cases would be the preferred anaesthetic.

References

1 *Riccardi VM.* Von Recklinghausen neurofibromatosis. N Engl J Med 1981; 305: 1617–27.

2 *Tapp E, Hickling RS.* Renal artery rupture in a pregnant woman with neurofibromatosis. J Pathol 1962, 97: 398–9.

3 *Ginsburg DS, Hernandez E, Johnson JWC.* Sarcoma complicating von Recklinghausen disease in pregnancy. Obstet Gynecol 1981; 58: 385–7.

4 *Brady DB, Bolan JC.* Neurofibromatosis and spontaneous hemothorax in pregnancy: two case reports. Obstet Gynecol 1984; 63: 355–75.

5 *Edwards JNT, Fooks M, Davey DA.* Neurofibromatosis and severe hypertension in pregnancy. Br J Obstet Gynaecol 1983; 90: 528–31.

6 *Morgan BM, Bulpitt CJ, Clifton P, Lewis PJ.* Analgesia and satisfaction in childbirth (the Queen Charlotte's 1000 Mother Survey). Lancet 1982; 2: 808–10.

7 *Stoelting RK, Dierdof SF, McCammon RL.* Anesthesia and Co-existing Disease, 2nd ed. New York: Churchill Livingstone, 1988: 347–8.

8 *Horyn G, Bourjeois-Dujols, Palaric JC, Giraud JR.* Maladie de Recklinghausen et complications vasculaires au cours de la grossesse. J Gynecol Obstet Biol Reprod 1988; 17: 641–5.

9 *Mallampati SR, Gatt SP, Gugino LD, et al.* A clinical sign to predict tracheal intubation: a prospective study. Can Anaesth Soc J 1985; 32: 429–34.

10 *Crozier W.* Upper airway obstruction in neurofibromatosis. Anaesthesia 1987; 42: 1209–11.

11 *Chang-Lo M.* Laryngeal involvement in von Recklinghausen's disease: a case report and review of the literature. Laryngoscope 1977; 87: 435–42.

12 *Fisher MM.* Anaesthetic difficulties in neurofibromatosis. Anaesthesia 1975; 30: 648–50.

13 *Humble RM.* Phaechromocytoma, neurofibromatosis and pregnancy. Anaesthesia 1967: 22; 296–303.

14 *Rubenstein AE, Wallace S, Aron AM, Penchazadeh G.* Neurological complications in 250 cases in neurofibromatosis. Ann Neurol 1984; 16: 133–4.

15 *Manser J.* Abnormal responses in von Recklinghausen's disease (Letter). Br J Anaesth 1970; 42: 183.

16 *Magbagbeola J.* Abnormal responses to muscle relaxants in a patient with von Recklinghausen's disease (Letter). Br J Anaesth 1970; 42: 710.

17 *Baraka A.* Myasthenic response to muscle relaxants in von Recklinghausen's disease. Br J Anaesth 1974; 46: 701–3.

18 *Magnin G, Philippon B, Chollat L, Soutoul JH.* Evaluation de l'irradiation pelvienne au cours des examens tomodensitométriques. Rev Fr Gynecol Obstet 1984; 79: 1–5.

19 *Ginsberg JS, Hirsh J, Rainbow AJ, Coates G.* Risks to the fetus of radiologic procedures used in the diagnosis of maternal venous thromboembolic disease. Thromb Haemost 1989; 61: 189–96.

REFERENCES (Crochetière)

20 *Naulty JS.* Epidural analgesia for labor. *In:* Norris MC (Ed.). Obstetric Anesthesia. Philadelphia: Lippincott, 1993: 319.

21 *Smith JJ.* Antenatal care – past, present and future (Editorial). Obstetric Anesthesia Digest 1994; 14: 47–8.

22 Deaths associated with anaesthesia. *In:* Report on Confidential Enquiries into Maternal Deaths in the United Kingdom, 1988–1990. London: HMSO, 1994; 80–96.

23 *Rocke DA, Murray WB, Rout CC, Gouws E.* Relative risk analysis of factors associated with difficult intubation in obstetric anesthesia. Anesthesiology 1992; 77: 67–73.

24 *Chervnak FA, McCullough LB.* Clinical guide to preventing ethical conflicts between pregnant women and their physicians. Am J Obstet Gynecol 1990; 162: 303–7.