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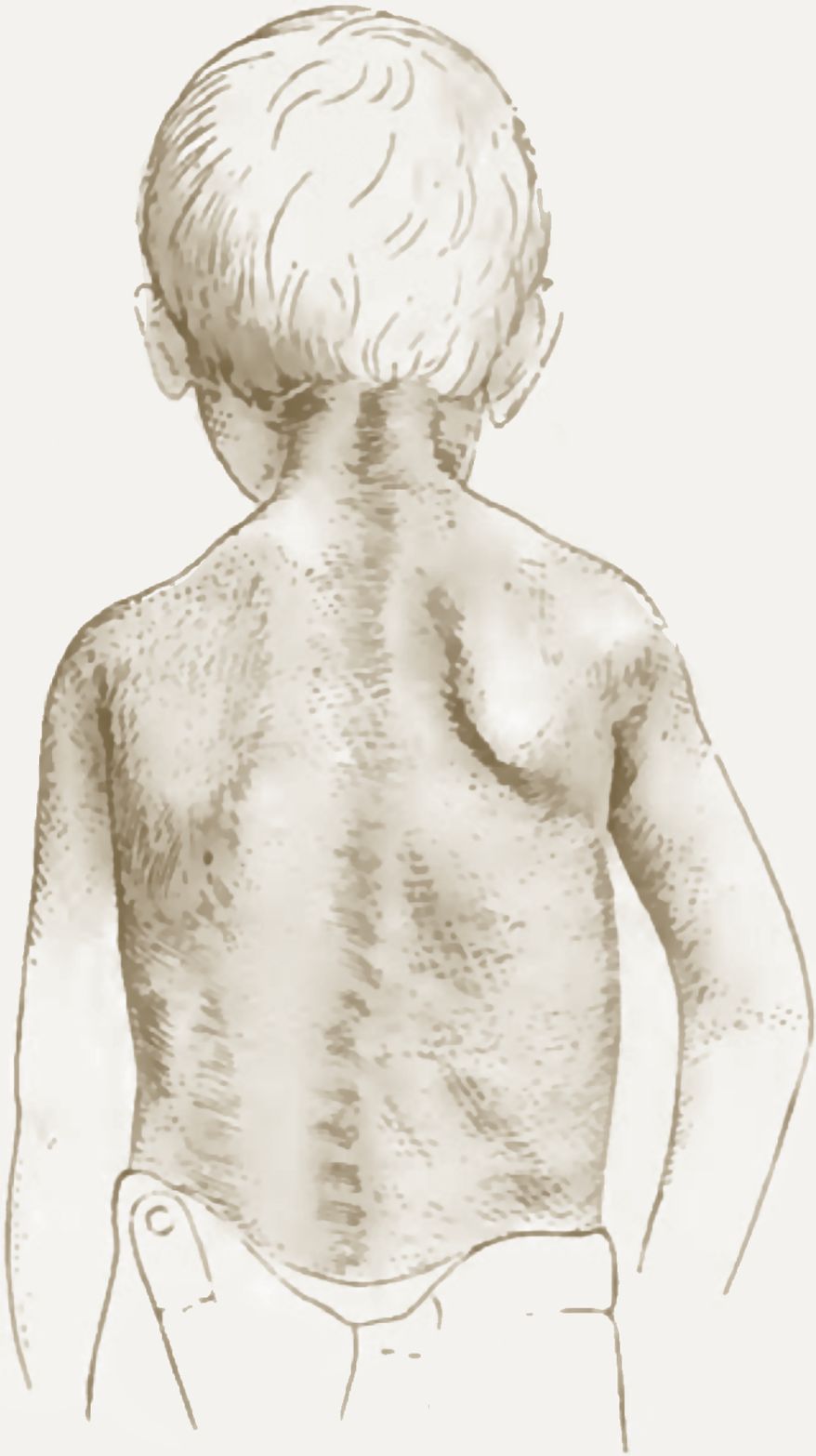

Fig. 53.

Part 3

Electrophysiological support for prognosis and diagnosis

Duchenne described in great detail four patients with an obstetric brachial plexus palsy, recognizing that their shoulder joint deformity was the result of a paralysis of the shoulder muscles. The reaction of muscles to direct application of electrical current was investigated thoroughly, making Duchenne one of the first electromyographers.

G.-B. Duchenne de Boulogne. De l'Électrisation localisée et de son application al la Pathologie et la Thérapeutique. 1855

Chapter 6

A review of electromyography in OBPL

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Abstract The few studies on prognosis of obstetric brachial plexus lesions that are not hampered by selection bias or a short follow-up suggest that functional impairment persists in 20-25% of cases, more than commonly thought. Electromyography (EMG), potentially useful for prognosis, is often considered of little value. Denervation in the first week of life has been interpreted as evidence of an antenatal lesion, but is the logical result of the short axonal length affected. EMG performed at close to the time of possible intervention (3 months) usually shows a discrepancy: motor unit potentials are seen in clinically paralyzed muscles. This can be explained in five ways: an overly pessimistic clinical examination; overestimation of EMG recruitment due to small muscle fibers; persistent fetal innervation; developmental apraxia; or misdirection, in which axons reach inappropriate muscles. Further research into the pathophysiology of obstetric brachial plexus lesions is needed to improve prognostication.

Obstetric brachial plexus lesions (OBPLs) are usually caused by traction to one brachial plexus.[18,46] As severity varies between neurapraxia, axonotmesis, neurotmesis, and root avulsion,[93] and the extent of injury varies between damage to one nerve or all roots, the impact of OBPL ranges from temporary functional impairment to a lifelong total paralysis of one arm.

OBPL is rare,[5] which may have contributed to a considerable uncertainty concerning key issues, treatment in particular.[46] Some investigators refrain from surgery completely, in view of the purportedly good chances of spontaneous recovery. Others advocate a "wait-and-see" approach,[63] reserving surgery for infants in whom spontaneous repair is deemed insufficient after a certain period, the length of which is itself a matter of debate.[15,26,50] This uncertainty is due to a lack of appropriately designed outcome studies, deplored in various editorials. [10,46,81] Another major difficulty is the lack of reliable indicators of prognosis. Electromyography (EMG) would seem an ideal tool for this purpose, but again reports are contradictory: some positive views have been presented,[20,90,91] but many investigators have expressed serious doubts about the value of EMG.[18,33,43,50,87,89] This controversy regarding the role of EMG in OBPL contrasts strongly with its clear-cut role in adult plexus lesions.

There appears to be only one study comparing the prognostic accuracy of clinical, radiologic, and EMG findings, suggesting the utility of the EMG, but the limited size and follow-up duration prohibit generalization of the conclusion.[105] There is often a striking discrepancy in OBPL between a clinical paralysis, on the one hand, and EMG findings suggesting functional innervation, on the other hand, by the presence of motor unit potentials (MUPs) in the absence of "denervation activity" (fibrillation potentials and/or positive sharp waves).[100,102,106]

This discrepancy probably underlies the perceived lack of utility of the EMG in OBPL. This review outlines major clinical features of OBPL and considerations regarding surgery, and presents a conventional interpretation of the EMG findings in OBPL. This is contrasted with other possible interpretations of the findings, which together urge a re-examination of the role of the EMG.

Clinical features of OBPL

Incidence and risk factors

Based on eight studies, OBPL has shown a mean incidence of 0.12% of births, with a range of 0.04-0.20%.[47] The most common delivery pattern concerns children presenting in the vertex position, where progression of the shoulder is blocked by the symphysis, causing traction of the brachial plexus (shoulder dystocia). Compared to routine delivery, dystocia doubles delivery forces.[3] The main risk factor for shoulder dystocia is macrosomia, which occurs in maternal diabetes. [51,104] Additional factors for the occurrence of OBPL include method of delivery,[25,61] multiparity,[104] and ethnic background.[30,104]

A less common delivery pattern concerns children, usually with low birthweight, born in a breech position.[23] This pattern carries a high risk of root avulsion.[23,98] In fact, it has been argued that breech presentation should always be followed by caesarean section,[36] although OBPL has been described after caesarean section.[44]

Patterns of damage

Four major patterns of injury can be distinguished, based on a craniocaudal spread of the lesions.[65] In group I, the Erb/ Duchenne pattern, C5 and C6 roots are affected exclusively; this pattern comprises about one half of patients.[11,92] In group II, comprising one third of cases, C5-C7 functions are impaired.[11,92] This type results in the typical "waiter's tip" posture: the shoulder is adducted and internally rotated; the elbow is extended; and the wrist and fingers are flexed (Fig. 1). In group III (15% of cases), some finger flexion remains present, but the arm is otherwise paralysed. [11,92] The group IV pattern involves a flail arm with Horner's sign due to severe damage to all roots.

The rare (Déjèrine-)Klumpke type of OBPL falls outside this pattern and involves isolated paralysis of the hand with Horner's sign.[2] This type is often associated with

Figure 1: **Three-month-old infant with OBPL**

Note the abnormal posture of the left arm. The shoulder is adducted and internally rotated, pointing to weakness of the deltoid and infraspinatus muscles (largely C5). The elbow is extended as a result of biceps weakness (largely C6). The wrist and fingers are flexed due to weakness of extensors (largely C7).

avulsion of the C8 and T1 roots. Associated injuries include phrenic nerve palsy, clavicular fracture, dislocation of the humerus, fractures about the shoulder girdle, facial nerve palsy, and torticollis.

Clinical examination

Examination is difficult and imprecise because of the lack of cooperation. Careful observation is a prerequisite. The observer should be aware of "trick" movements. For instance, the elbow may be flexed through action of the wrist extensors, mimicking a functional biceps. Another is seen when a child, lying on his back, flings an arm upward with the pectoral muscles; the elbow is then flexed through the effect of gravity, imitating a functional biceps muscle. Systematic examination starts with the passive range of joint motion, allowing contractures to be recognized. Second, active joint excursions should be examined in different positions to assess muscle strength, taking effects of gravity into account.[15] As in adults, strength can be quantified with the Medical Research Council (MRC) grading system,[86] but differentiation between grades 3 (motion against gravity), 4 (weakness), and 5 (normal strength) is often difficult. Some investigators have proposed combining all levels, with overcoming the effects of gravity as grade 3.[95] At the other end of the spectrum, it is conceivable that limited movements, corresponding to MRC grades 1 and 2, may be missed entirely. Although such levels are functionally irrelevant, they are important, as their presence argues against a total loss of neural continuity and thus against immediate surgery.

Clarke introduced a scoring system in which functional movements of the hand and arm are given points, resulting in a sum score, which serves as a predictor for recovery and as an indicator for nerve surgery.[15]

Prognosis

Many reviews have indicated that the general prognosis of OBPL is good, meaning that spontaneous recovery will occur in over 90% of cases.[30,32,37,50,67,96] However, we believe that prognosis may not be as good as believed for the following three reasons.

The first reason lies in assessing the functional end stage. As noted earlier, the neurological examination is imprecise. Besides a lack of cooperation, immature motor control may be to blame: less severe lesions may only become apparent when motor control becomes sufficiently subtle to require the full range of movement. Sjöberg et al. stressed that children may feel more inadequate as demands increase with age.[88] In adults, spontaneous recovery of nerve injuries is usually thought to reach an end stage 2 years after injury; as a similar period may be applicable in children, earlier assessments are probably inadequate.

A unique problem in OBPL is that the affected arm may grow less than the normal one. This contributes to the functional handicap, but cannot be assessed at an early age. Furthermore, persistent palsy is also associated with psychosocial problems, not yet present in infants.[7] Finally, co-contraction (or "synkinesis") has been shown to affect the functional end stage in older children and adults with OBPL. This refers to activation of antagonistic muscles or muscles not normally involved in the intended

movement, along with contraction of an agonist.[16,35,39,73,75] Recurring patterns of co-contraction in OBPL include: activation of the triceps when elbow flexion is intended [8,16,39,73]; activation of shoulder adductors when abduction is intended[16,35]; and activation of the "trumpet sign," which consists of activation of shoulder abductors when elbow flexion is intended, causing a posture resembling that of holding a trumpet before the mouth.[16,75] Co-contraction can seriously affect arm function; for example, in one study of 25 cases, shoulder abduction was impaired because of weakness only in 5 cases, adduction co-contraction in 15, and combined effects in the remaining 5.[35] In other words, co-contraction was more important than simple weakness. (Recent therapeutic interventions aim to abolish this counterproductive effect of nerve regeneration with botulinum toxin[73] or muscle transposition.[16]) Together, these factors suggest that a functional end stage in OBPL can only be assessed reliably in children of about 4 years or older. Even then, specialized predefined assessment protocols are needed, but these have been used only rarely.[6,58,94] An example of a protocol emphasizing functioning in daily life is that described by Sundholm et al.[94] The absence of such defined test protocols in many earlier studies casts doubts on their reliability. Studies in which severity of OBPL was assessed at an early age are likely to have erred on the optimistic side.

The second reason to doubt estimates of prognosis lies in case ascertainment; that is, studies based on referral to specialized centres or on hospital records may be biased in unknown directions. Only population-based studies with complete case ascertainment are likely to be able to avoid this error, and there are only few such studies.[5,30,42,61,88]

The third reason is that surgery was used in many studies. As the decision to operate was usually based on an early assessment of severity, such studies can clearly not be used to disentangle prognosis and the reliability of its assessment. In view of these considerations, an ideal study on the natural history of OBPL would consist of complete ascertainment of all cases in a population over a certain period; there would be no surgical intervention, and severity would be assessed at an age of at least 4 years using a specific, standardized protocol. Ideally, children should also be assessed after birth and at about 3 months of age, to determine how well the functional end stage can be predicted at an early age. There are no studies that fulfil all these criteria, but two Swedish studies come close. The first was based on complete case ascertainment in the city of Malmö over a 10-year period; surgery was not performed, and outcome was assessed at ages of 3-11 years.[88] The investigators reported persistent palsy in 12 of 48 (25%) cases, with "considerable reduction in arm function." The second concerned complete ascertainment in the county of Skaraborg, and assessment with a predefined program at the ages of 4-14 years.[5] Of 52 cases, 11 (22%) had severe stationary impairment of arm-hand function.

We conclude that estimates of prognosis of OBPL are based largely on insufficient assessment, and that one fifth to one quarter of cases may result in significant impairment.

Decisions regarding surgery

Discussions regarding surgery currently focus on whether it is justified at all, and, if so, when it should be performed. The first question depends, as in adults, on the presence of neurotmesis or root avulsion, as spontaneous recovery is then impossible. As this is most difficult to establish in OBPL of groups I and II, discussions regarding surgery are most relevant in these types. If the presence of such lesions could be assessed unequivocally, the problem would largely be solved. Unfortunately, neither clinical examination nor any additional test, including ultrasound or radiological imaging methods, can presently answer this question with sufficient reliability. It is generally agreed upon that results of nerve surgery are better in the first year of life than later, so unequivocal neurotmesis or root avulsion presents a strong case for surgery in the first months of life. As more roots are avulsed, fewer functions can be regained through surgery. Nerve transfers can then be applied in which nerves from outside the plexus are connected to elements within the plexus (e.g., intercostal-to-musculocutaneous nerve transfer).[57]

Unfortunately, accurate assessment of severity at 1-2 months of age is not possible, so decisions are often postponed in the hope of spontaneous recovery. If this occurs, surgery with its associated risks is justifiably avoided, but if it does not the results of surgery may be worse than if it had been carried out earlier. Some investigators advocate palliative surgery, such as tendon transfer,[92] as a useful alternative to primary nerve surgery. This opinion is not generally accepted, as others state that the results of tendon transfers in OBPL are inferior to those of good nerve regeneration.[9]

There is also no consensus regarding the guidelines for timing decisions. Two main points of view can be distinguished.[33] One favours nerve surgery at about 3 months of age, based on the absence of biceps contraction; the other calls for surgery at 9 months of age, based on the score of the Toronto Functional Assessment scale.[15,95] Although both methods focus on function, there is an important difference in the way infants may achieve function.

In the "3 months/biceps" approach, elbow flexion must be achieved through the biceps muscle, whereas it may be based on "trick" movements in the other approach. Although some infants develop such tricks, others with comparable lesions do not. Although elbow flexion may be present in both mechanisms, there are consequences for arm motility as a whole. There is hardly any information on long-term effects, so there is no answer to the question of whether both approaches are equally valid.

In short, there are insufficient data to feel confident when deciding on whom to operate and when. Comprehensive outcome studies on prognosis are needed before these questions can be answered.

EMG findings in OBPL: the problem

EMG in OBPL presents two problems, both of which are related to the age of the infant.

EMG in the first week of life

Several investigators have noticed early denervation activity in the first few weeks of life in OBPL, and concluded that the lesion must have occurred before birth,[19,49,69-71] although some did attribute it to birth trauma.[62] This conclusion was explicitly or implicitly based on the interval of 10-14 days that usually elapses between nerve injury and the appearance of denervation activity. Similar conclusions were drawn to explain denervation activity in cases of peroneal neuropathy[80] or radial nerve palsies in newborn infants.[74] If a lesion occurs well before birth, obstetric procedures cannot be held responsible, with legal and financial consequences. Some investigators have advocated EMG in the first week of life for this particular reason.[68]

EMG at 3 months of age

Decisions regarding surgery are often made at around 3 months of age, as the need for diagnostic procedures is greatest at that time. A common finding is that EMG contradicts the clinical examination.[99,103,106] Clinically, there may be complete paralysis of shoulder abduction and elbow flexion. In an adult, one would then expect EMG to show profuse denervation activity and absence of motor unit potentials (MUPs) in the biceps and deltoid muscles. In contrast, in a child with OBPL, MUPs are present, and denervation activity is absent. Recruitment patterns usually cannot be ascertained with confidence, but observation suggests the firing pattern to be sufficiently "full" to allow at least some visible movement. Similar findings were reported as early as 1965.[106] This discrepancy does not occur in all cases. In infants with paresis instead of paralysis, MUPs without denervation activity are expected. In some cases with paralysis, the expected picture of denervation activity without MUPs does occur, but these patterns occur less often than the contradictory findings discussed previously.

Paradiso et al. described 100 EMG studies in 78 children with OBPL of Erb's type, aged between 5 days and 15 months.[70] Denervation activity was invariably present from day 10 through day 60, and was last seen during week 19. Abnormal MUPs (high-amplitude, long-duration, polyphasic or small polyphasic MUPs) were first seen at week 9, and were present in all patients from week 13 on.

These data show that EMG develops in OBPL as it does in adults; that is, denervation appears, and later disappears, to make way for altered MUPs. But the resemblance with adults ends there: How can denervation activity appear in the first week of life, too early to attribute it to birth trauma? Why is it so conspicuously absent in clinically paralysed muscles at 3 months of age? How can MUPs be present in these same muscles at that age? How can the presence of MUPs be reconciled with the absence of any recognizable movement?

EMG findings: possible answers

Quick appearance of denervation activity

Many textbooks indicate that denervation activity starts 10-14 days after axonotmesis, but they fail to stress that the timing of onset of denervation depends on both the

severity of the nerve lesion and the length of the nerve between muscle and the site of injury. In small animals, denervation activity starts earlier. In rats, denervation activity has been shown to begin 42 hours[83] or 90 hours[54] after injury. Other rat studies have shown that denervation begins a day earlier after complete than after partial lesions.[4]

Short nerve lengths probably cause rapid denervation because the remaining nerve segment is quickly emptied of trophic factors. As the nerve length from the plexus to the biceps is about 300-400 mm for an adult, and 100 mm for an infant, emptying may occur three or four times earlier. The diameter of motor axons of infants is about 5 µm in infants and 8 µm in adults,[85] making the cross-section in infants about 2.5 times smaller. Together, these two factors might cause draining of the axon to be complete 7.5-10 times sooner than in adults. Compared with the 10-14-day adult period until appearance of denervation activity, a period of 1-2 days, or even less, is to be expected in infants. One study in pigs was designed to investigate the time of onset of denervation in OBPL, and showed that denervation after nerve section in the deltoid muscle occurred after 24 h in 2-day-old piglets (this was the first EMG after trauma) and after 5 or 8 days in two adult pigs.[28]

The 10-14-day interval between lesion and onset of denervation is thus wholly inappropriate in infants. Serial data on very early EMGs are lacking in OBPL, so it is not known when denervation activity due to a lesion at birth may first become visible. Because a period of 1-2 days is highly probable, early denervation activity may not be used to prove putative intrauterine damage. In fact, the vast majority of cases with early denervation are probably simply due to an injury during birth. An earlier lesion would also require the presence of devastating forces in utero before the onset of labour. Although we are not qualified to address this problem, we tend to agree with Royden Jones that it is hard to think of a mechanism with enough force to avulse nerve roots in utero.[79]

This does not mean that intrauterine plexus palsy is wholly impossible, but other evidence should be sought to prove its presence or absence. Antenatal plexus injury has been implicated in an extraordinary case of a child born with a smaller, atrophied, flaccid arm due to a bicornuate uterus.[19] In very rare cases, plexus lesions in infants may be due to tumors. [1] The presence of clearly altered MUP morphology early in life may also be seen as evidence of a prenatal lesion,[70] although the unknown sensitivity and specificity of this finding in infants cast doubts on its utility. Note that we do not state that intrauterine plexus or nerve lesions [80] are impossible, but that the presence of denervation activity indicates that the lesion occurred about 1 day prior to investigation, or earlier.

Discussions in the obstetric literature appear to revolve around the issue whether the forces acting on the brachial plexus were exerted by obstetricians (creating an opening for possible lawsuits) or the result of unavoidable pressures during birth.[24,44,45] Mathematical models [29] or the presence of OBPL after caesarean section and in births without obstetric help [44] have shown that the excess force need not be applied by obstetricians. Whatever the mechanism, the presence of early denervation is of no value to either the plaintiff or the defence in a court of law.

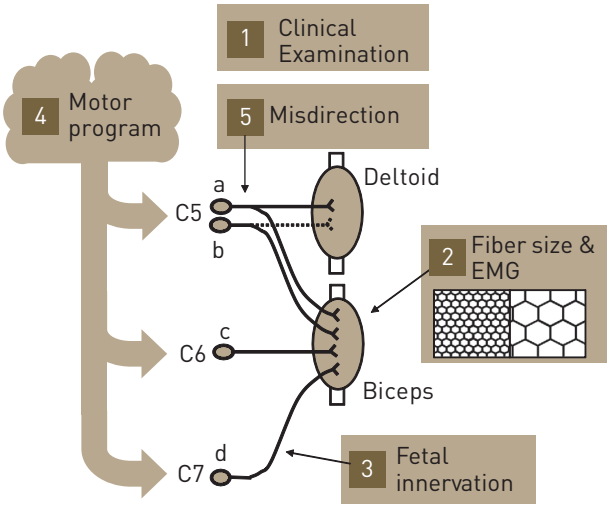
Disappearance of denervation activity

A disappearance of denervation activity between days 10 and 60 of age[70] shows that muscle fibres are literally no longer denervated. Still, the process of muscle denervation may differ from that in adult muscles. Muscle biopsies in two cases of denervation in infants showed small round fibres, similar to those seen in Werdnig-Hoffmann disease.[27] One biopsy was taken at 8 weeks in a case of OBPL, and the other taken in the third month of life following an iatrogenic lesion during the first week of life. The investigators concluded that the process of muscle denervation was age-dependent.[27]

Presence of apparently inactive MUPs

The problem can be rephrased as follows: If the presence of MUPs at 3 months of age indicates a functional contact with axons, why does this not result in observable movement? Five different reasons may be given, as discussed in what follows (Fig. 2). Co-contraction can also be explained through several of the mechanisms involved.

Figure 2: **Explanations for apparently inactive MUPs in OBPL**



This schematic representation of biceps and deltoid innervation shows various explanations for apparently inactive MUPs in the EMG of the biceps muscle. Full explanations are given in the text. Severe paresis may be mistaken for a complete paralysis due to lack of cooperation during clinical examination (1). The EMG may be interpreted as "too optimistic" (2); as muscle fibre diameter is 3.3 times smaller in infants, 11 times more fibres are present in the uptake area of the needle (left panel) than in adults (right panel). Persistent foetal "luxury" innervation of the biceps muscle through the "wrong" root (C7) may persist after birth (3). Abnormal development of motor programs may cause an improper development of agonist/ antagonist firing patterns (4). Finally, incorrect outgrowth may cause inappropriate firing (5). In complex misdirection (a), a nerve fibre from C5 meant for the deltoid also gives off a branch to the biceps. The two parts of this motor unit in the two muscles will respond to abduction commands. In simple misdirection (b), a C5 axon meant for the deltoid ends up in the biceps without branching. Fibre (c) runs from C6 to the biceps and represents normal innervation.

1. Inadequacy of the clinical examination

As indicated previously, neurological examination of infants is necessarily limited. Observing movements for several minutes with an EMG needle in place has shown that bursts of EMG activity may be associated with very slight movements that would not have been noticed without a simultaneous EMG. We fear that the clinical examination may be overly pessimistic in severe injuries, suggesting that there is no neuronal continuity to the muscle when it is, in fact, present. The "inactive MUPs" then correspond to strength of MRC grades 1 or 2.

2. Overestimation of the number of MUPs

The number of muscle fibres does not change during life,[52] but their size does; that is, fibres are smaller in infants. [12,13,66,101] Muscle fibre diameter increases from 17 µm in 3-month-old infants[13] to 57 µm in adults,[12] or 3.35-fold. The diameter of deltoid muscle fibres increases 3.29-fold, from 15.2 µm to about 50 µm.[66] The cross-sectional area of infants' fibres is therefore about 11-fold smaller.[99] Provided that the number of muscle fibres per motor unit stays the same during life, 11-fold more motor units can be recorded for the same EMG needle uptake area. The amplitude of a single muscle fibre potential will be lower, as this too depends on fibre size, but its effect may be partially compensated by the smaller distances between muscle fibers.[100] Data on MUPs in infants have shown that their amplitude is lower and duration shorter in infants of 3 months as compared with adults. In that study, the shorter duration in infants was attributed to the narrow width of the endplate zone (i.e., short distances between muscle fibres).[82] The uptake area of EMG needles depends on type,[49] but as long as similar-type needles are used in adults as in infants, the estimate of the number of active motor units may be a gross overestimate in infants, leading to an overly optimistic view.[99]

3. Luxury innervation

Vredeveld et al. found that intraoperative stimulation of the C7 root in infants with OBPL evoked elbow flexion and shoulder abduction in infants with Erb's type, whereas stimulation of C5 or C6 roots did not.[102,103] This would suggest that the biceps and deltoid muscles received innervation through the C7 root, which is normally not the case. The explanation was sought in "luxury innervation," meaning that muscle fibres are equipped with more than one neuromuscular synapse during early stages of foetal development. [40] For this explanation to be correct, polyneuronal innervation must still be present at birth, but the evidence for this is conflicting. One study reported that the transition from polyneuronal to mononeuronal innervation takes place well before birth, at between 16 and 25 weeks of development.[40] Another investigation stated that polyneuronal innervation is still present in the human psoas muscle until week 12 after term age.[41] It is not yet known whether the multiple nerve endings on a muscle fibre derive from one axon, multiple axons of the same root, or from multiple segments. According to this concept, normally superfluous C7 neurons are not lost in OBPL, because they do not compete with the absent C5 or C6 neurons; thus, they have little functional relevance because of a central failure to drive them correctly.

4. Central motor disorders

OBPL not only affects motor output, but also sensory input. Many neurologic systems exhibit a critical period for formation, dependent on afferent impulses. The deafferentation of OBPL may inhibit the development of normal motor control, which can remain abnormal even if later peripheral nerve repair partially ameliorates the peripheral part of the problem.[59,99,106] In a rabbit study designed to address this problem, functional impairment was larger for lesions sustained at birth than for lesions later in life, although peripheral regeneration was shown to occur at all ages.[106] The mean number of motor units in biceps and thenar muscles was approximately 50% that of control arms in a study in children with OBPL with a minimum age of 4 years.[84] MUP amplitudes were increased; the pattern was therefore that of severe loss of motor units of long standing. Eight children had normal numbers of motor units in the biceps muscle, in contrast to the significant upper arm and shoulder problems. Agnosia or apraxia was hypothesized.[84] A follow-up study revealed that voluntary elbow flexion force in the affected arms was considerably lower than the force exerted by electrical stimulation of the muscles,[14] whereas forces were similar in control subjects. In short, subjects with OBPL had fewer biceps motor units than control subjects, and they were unable to recruit all of those they did have.[14] "Developmental apraxia"[14] was suggested to explain why some motor units were not put to work through voluntary commands. We have repeatedly observed that children with OBPL will flex their elbows while picking up a ball, but will "forget the arm" during running or when they otherwise neglect to focus on the use of the arm. This has also been mentioned by others.[94] This suggests that use of the arm depends on the requested movement pattern (i.e., on central patterns).

It is conceivable that disordered motor programs do not merely result in a lack of firing at the right moment, but also in firing when this is not required. This mechanism has been implicated in co-contraction in OBPL,[75] and may explain "inactive MUPs."

5. Abnormal nerve branching

Motor axons growing toward the periphery after lesion need not regain their original target. In fact, there appears to be no guiding principle in outgrowth after axonal section, and thus reinnervation appears to be an essentially random process. Axons may reach agonistic, antagonistic, or unrelated muscles.[31] Because motor neurons still fire in response to their original driving patterns, flexors may be activated together with extensors, resulting in severe movement disturbances. [31] In "complex misdirection," axons split into various branches that may end up in various muscles or even in cutaneous nerves (Fig. 3).[8,17,75-78] Roth stimulated ulnar and median nerves, motor points of various muscles, and sensory nerves in the fingers, and recorded resultant MUPs ("heterogeneous axon reflexes") from a large number of muscles in 16 cases of OBPL, aged 1-47 years.[75-78] These were found in all 16 cases (up to 100 in one patient). Out of 618 searches for a communication between two muscles, or between a muscle and an inappropriate nerve, no less than 38% resulted in such heterogeneous axon reflexes. Abnormal sensory-muscular communications were found in 10 of 14 subjects.[75] Similar sensory-motor communications were, almost without exception, also found in

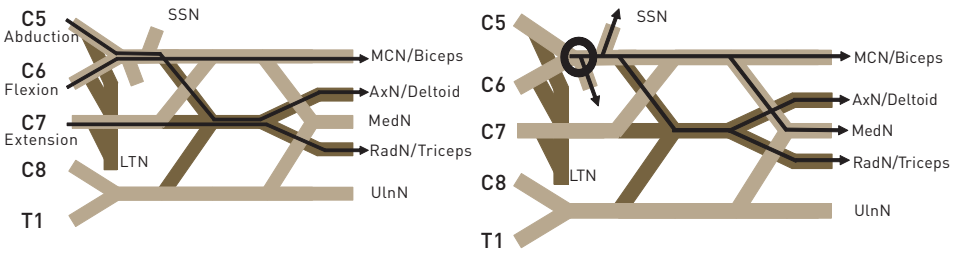
adults with previously sutured peripheral nerves.[64] In "simple misdirection" the axon does not split, so the motor unit forms in its entirety in the wrong muscle.[21]

Both types of misdirection can result in co-contraction of various muscles, and can explain "inactive MUPs"; instead these belong in another muscle. In adults, misdirection can be tested by asking subjects to contract muscles other than the one the needle is in, but this cannot be done in infants. Routine experience in adult plexus lesions has shown that misdirection is a regular occurrence. Still, co-contraction in adult plexus lesions is never as severe a problem as it is in older children with OBPL. One possible explanation is that axons in infants are more likely to split than adult axons. There is animal evidence that nerve regeneration differs between adults and infants, as shown by the longer persistence of unmyelinated axons in rats,[97] increased axonal branching in neonatal rats,[38] earlier axonal degeneration and recovery in foetal sheep,[53] or increased regeneration capability in young sheep.[22] It is therefore conceivable that the tendency for axonal branching differs between infants and adults.

EMG findings: what is the solution?

The quick appearance of denervation activity is simply a consequence of infants' axons being shorter and thinner. Its quick disappearance and the emergence of MUPs point to regeneration. Nevertheless, it remains unknown as to why these MUPs, at around 3 months, appear to have no function. The five possible explanations given previously are not mutually exclusive, as none renders any of the others impossible.

Figure 3: **Nerve outgrowth in OBPL**



a) Normal innervation pattern. Motor commands for shoulder abduction project on C5 motor neurons that innervate the deltoid muscle; likewise, commands for elbow flexion reach the biceps through the C6 root, and elbow extension is effected through the C7 root and the triceps muscle (in reality, muscles are innervated through more than one root, and more than one muscle contributes to each function). b) The situation after severe proximal axonotmesis in the superior trunk (circle). Outgrowing axons may or may not split, and can grow into all available pathways downstream from the superior trunk. Motor commands may then reach unintended muscles. For instance, axons destined to effect elbow flexion may now innervate the biceps, but also its antagonist (triceps) or the deltoid muscle. SSN, suprascapular nerve; LTN, long thoracic nerve; MCN, musculocutaneous nerve; AxN, axillary nerve; MedN, median nerve; RadN, radial nerve; UlnN, ulnar nerve; Biceps, biceps muscle; Deltoid, deltoid muscle; Triceps, triceps muscle

It would be surprising if the central nervous system were not involved at all, although this might take two completely different forms. The first is a permanent loss of function due to a closed window of opportunity to develop a motor program, as suggested earlier.[14,84] The other rests on the finding that central nervous system plasticity occurs in adult plexus lesions [55-57]; the young nervous system appears capable of surprising amounts of plasticity and functional repair, so a central "solution," involving recruiting muscles for trick movements, is not unreasonable.

Axonal misdirection has been shown to exist in older children with OBPL, and, as branching is part of nerve regeneration, it is highly likely that branching is present at 3 months of age. Axonal branching can explain why direct muscle stimulation was shown to result in more force than voluntary activation.[14] In that study, voluntary activation of the biceps did not excite all motor units in it, as some "belonged" to other motor programs. However, in response to criticism along these lines,[72] the existence of "heterogeneous axon reflexes" was doubted, because sensory-motor connections could have been due to normal cutaneous reflexes.[59] There is no need, however, to present these theories as conflicting; extensive peripheral "cross-wiring" would present the central nervous system with conflicting feedback, as a command for elbow flexion would be followed by feedback from flexors, abductors, or extensors, or some combination of these muscles. The inability to learn to activate muscles selectively might seriously disrupt normal agonist-antagonist programming. As noted earlier, the other explanations may also be true. In particular, it would be very surprising if the 11-fold tighter packing of muscle fibres in a muscle does not affect the interpretation of the EMG recruitment pattern to some degree.

Conclusion and prospects

Treatment and assessment of prognosis in OBPL suffer from a lack of methodologically sound evidence. OBPL presents a number of specific problems that affect the assessment of prognosis; we are currently performing a systematic review of prognosis that takes these factors into account. The EMG might be very useful for prognosis, but pathophysiological and longitudinal studies are needed to redefine when it should be performed and which parameters are useful for prognosis. Electromyographers and surgeons should realize that the presence of MUPs in a clinically paralytic muscle indeed indicates that there is continuity from spinal cord to muscle, and should be aware that factors other than mere continuity can also be relevant to regain useful function of the arm. This is not an academic question, as surgical efforts are largely aimed at restoring neural continuity. We therefore urge researchers not to restrict their focus to the restoration of continuity, but rather to widen their attention to encompass all factors affecting restoration of function. Focus points for such research should be the extent of axonal branching and the selectivity of the central drive to various muscles in obstetric and adult plexus lesions. Such knowledge will improve not just the utility of the EMG, but also in understanding the nature of nerve repair and motor control in infants. Ultimately, this may improve the treatment of this complex disorder, which will affect these individuals for their entire lives.

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