

# UNUSUAL BUT FREQUENT ETIOLOGY OF AN ACUTE PERINATAL DISTRESS IN TWIN PREGNANCIES

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*This paper aims at drawing the attention of obstetricians and pediatricians to a peculiar syndrome: a neonatal emergency without any preliminary signs of fetal distress. This emergency can be predicted before birth. The uterine content film obtained, when a twin pregnancy or an abnormal presentation is suspected, shows an extreme and permanent fetal malposition as early as the 6th month of gestation: the head is forcibly overextended, embedded in the spinal abnormal concavity. Its severity and duration entail a cervical elongation and a temporary atonia of inspiratory muscles. Respiratory autonomy is impossible at birth. A prolonged artificial ventilation should be organized and used immediately at birth to prevent death or any risk of transient cerebral anoxia. The acute neonatal neurological syndrome is only due to a purely peripheral motor and transitory impairment, and it will have no consequences.*

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Among the numerous examples of neonatal pathology, I chose to describe a particular one for three reasons: (1) it concerns both obstetricians and pediatricians; (2) it contains several practical lessons; (3) it is relatively frequent in twin pregnancies but has received little attention, chiefly with the survivor newborns.

This pathology was a slow discovery; the three following stories of babies submitted to a long resuscitation at birth will explain the slow progressive advance of our work. Whenever a baby presents an acute distress with severe and lasting neurological symptoms, it is natural to be immediately afraid that a cerebral damage will heavily bear on the child's future. However, given a supposedly similar acute accident, the far sequelae can be quite different from one child to another. Why is it so? Is it sometimes possible to predict the evolution, thus allowing to avoid irretrievable consequences by timely adopting the proper policy?

*The first baby*, to be such source of perplexity in relation with the etiology of its symptoms, was born from parents who were first cousins, born themselves from consanguineous parents. The baby was the product of a fourth pregnancy; the first three children all died within the first months after birth with severe encephalopathies. The geneticist said that the caryotype was normal, but a probable recessive trait spared very little chance for a normal child. The baby was born through Cesarean section, but needed a stormy resuscitation and was finally admitted to our Intensive Care Unit.

Throughout the first week of life, the neurological status remained alarming, in spite of the progressive clearing of the respiratory distress. The usual posture was very abnormal, with a major opisthotonos; the head was sagging backwards and totally dropping. However, around the 10th day of life, some sensory responses appeared. At 6 weeks of age, the motor anomalies persisted, but the developing psycho-affectivity was good; the head was still sagging, but the quality of smile was encouraging. At 3 months of age, an extreme discrepancy became obvious between the normal psycho-affective evolution and the



Fig. 1. Uterine content film at the 6th month of gestation. Twins. The second twin's definite malposition is already set: the head is overextended, with a deep cervical posterior concavity, as seen on the upper and left side.

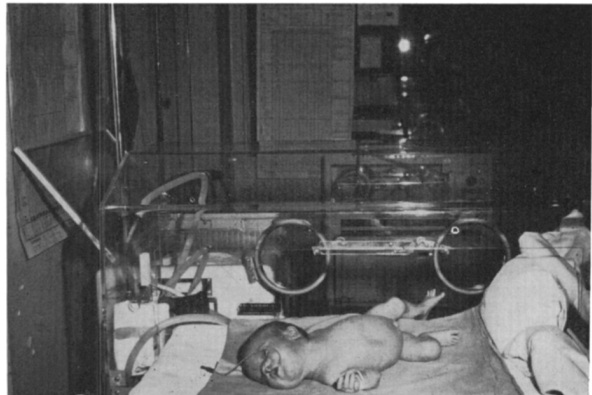


Fig. 2. In the baby born near term (three months after the film shown in Fig. 1), the permanent posture faithfully reproduces the fetal wrong position: the neck overextension and the atonia of inspiratory muscles account for the temporary absent respiratory autonomy through an exclusively peripheral motor impairment.

motor impairment: not the least attempt of correct head posture was achieved; when lying prone, the child was unable to rotate his head laterally and free his nose to breathe. What kind of future could be expected? Against our apprehensions, the motor anomalies disappeared around one year of age. Therefore, in spite of the family and neonatal history, in spite of the long resuscitation and lasting neurological signs, the child was unexpectedly normal. What had taken place?

*The second story* was going to prepare us the way to understanding. This baby was a second twin, born through breech delivery. After a long resuscitation, nonspontaneous respiration was obtained during the first days of life, and the baby was maintained on artificial intermittent positive pressure ventilation with a tracheal tube. A triple cord around the neck was supposed to account for those difficulties. However, when the acute episode was over, it was impossible to explain the irreducible "horn" posture. Furthermore, were the cord around the neck responsible for the lasting initial respiratory distress, some signs of cerebral damage would have been likely to appear. On the contrary, the psycho-affective development remained excellent; but the motor anomaly was still present at one year of age. The whole picture favored a peripheral damage more than a brain damage. The final hypothesis evoked by the "horn" posture was a possible fetal malposition in utero with possible cervical medulla elongation.

When *the third baby* was admitted to the Intensive Care Unit with a similar perinatal history and a strictly parallel evolution, we asked whether the pregnancy had been checked radiologically. By chance, a film was available, and it showed the wrong fetal posture that had been suspected and held responsible for the tonus anomalies observed at birth (Fig. 1). In utero, the fetal head was overextended backwards and embedded along the cervical backbone. This was the exact posture of the baby in its incubator yet at the 10th day of age (Fig. 2). Such an abnormal posture still was the comfortable one for the infant. The

clinical neurologic examination noticed that the head was absolutely dangling; when the infant lying supine was lifted up to the sitting position, the head remained dropping, due to the injury of the neck flexor muscles. When the sitting position was maintained, the infant was unable to straighten his head up; in our experience, the absence of this reflex usually represents a major neurological symptom, but in the present case, it was only the result of the hypotonia of the neck extensors. The truly peripheral and localized nature of the anomaly was confirmed by the following fact: contrary to expectations, all other straightening reactions were perfect. Finally, the good Moro reflex and the excellent vigilance demonstrated the soundness of the central nervous system. It therefore was not surprising that, in spite of the perinatal troubles, a fine quality of alertness was already observed at as soon as 10 days of age, and it constituted a heartening foresight.

Trying to solve the problem offered by our first such case, and particularly the underlying cause of the respiratory distress, an inquiry was done in search of a possible film; indeed, due to the mother's stormy history, the uterine content had been checked: it showed the same wrong fetal posture which had led to the same syndrome with the same unexpected issue.

Therefore, in spite of the heavy suspicion of cerebral damage being responsible for the neurological and respiratory difficulties at birth, in the three cases it was indeed a peripheral motor impairment, localized within the neck area. The head had been jammed into overextension early in pregnancy. The lasting duration of the malposition had entailed a medulla elongation and atonia at level of the neck and upper body is due to extended roots of the nerves emerging from the cervical medulla. The inspiration muscles being transiently inactive, the first breath could not be established, no cry was possible, and autonomous respiration was delayed.

This simple syndrome is understandable when the etiology is known, and it has practical implications: the severe respiratory distress, so harmful for the immediate life prognosis and the far intellectual prognosis, can be foreseen and therefore avoided. When a twin pregnancy, or a breech or forehead delivery are suspected, a radiological film of the uterine content is often obtained, to make the decision of vaginal delivery or Cesarean section. This very film can show the malposition and it then implies that before delivery everything be organized for an immediate intubation and a prolonged artificial ventilation, since the respiratory functions are likely to be delayed.

The obstetrician can predict the occurrence of difficulties from three criteria: (1) the *intensity* of the wrong fetal posture, involving the cervical and dorsal backbone; (2) the *long duration* of the malposition: in our cases, the uterine-content radiological film showed that the malposition was already set at the 6th month of gestation, but deliveries occurred at term or near term (these two points separate the syndrome from a simple "deflected head", which occurs late in pregnancy, is temporary, localized, less severe, and bears little consequences on the respiratory functions); (3) the *long resuscitation at birth* is quite unexpected when no prenatal diagnosis has been made, since it occurs as a single finding: no dynamic dystocia and no signs of fetal distress herald the birth of the severely affected infant.

The syndrome is an illustration to the fact that a striking neonatal symptomatology can offer a misleading interpretation as a birth trauma and a central, perhaps irreversible, brain damage. The etiology may in fact be transient and purely peripheral. The latter eventuality makes it mandatory to achieve the utmost care to save a newborn's life and future. There is no hurry to give up.

A severe and durable neurological manifestation is not needs the unescapable consequence of a brain damage. A striking clinical picture may be temporary. The interest of

semiologic neurological investigation lies in its ability to separate the simple associated symptoms and the major signs, the transient anomalies and the definitive ones; its other aptitude is to point out the true etiology that will have to be verified.

Those three difficult cases also are an example of the necessity for a better junction between obstetricians and pediatricians. Obstetrical information often undergoes a fairly incomplete transmission to pediatricians. Under such circumstances, the uterine content film would have quickly enlightened us about the etiology and, therefore, the correct emergency treatment of this peculiar neurological syndrome.

Neonatal pathology can offer a difficult approach because some seemingly obvious causal relationships are not necessarily responsible of acute accidents. Our ignorance is a basic reason to fight for any jeopardized neonate, to increase our current knowledge of the ultimate etiology, which may be avoided when timely recognized.