CASE REPORT

Exsanguinating Cephalhematoma:

A Report of Two Cases

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WITHIN a week or two of reading Van der Horst's[1] description of exsanguinating cephalhematoma in African newborns, two clinical cases of this disorder were seen at the War Memorial Children's Hospital, London, Ontario, and are described in the following report in the expectation that others may encounter similar cases.

CASE 1.—Baby "S" was born in August 1963 to an epileptic mother who was well controlled on primidone, mephobarbital and acetazolamide. The pregnancy was normal and the delivery at term was by the vertex with low forceps and an episiotomy. The infant's birth weight was 6 lb. 10 oz. No vitamin K was administered to mother or baby. Even in the delivery room, subcutaneous extravasation of blood was noted on the baby's legs. Over the next 24 hours many bruises appeared on the trunk, and the head circumference rapidly increased. The baby became lethargic but irritable, and later appeared obviously very pale, wizened and collapsed. At 40 hours of age she was referred to the War Memorial Children's Hospital, London, Ontario, from the Strathroy General Hospital.

On admission, the baby was shocked and exsanguinated. She was unresponsive to stimuli, made very feeble sucking efforts only, and the Moro reflex was weak. There were many bruises on the trunk and limbs, but no purpuric spots or petechiae. The head presented a remarkable appearance strikingly similar to that illustrated in Figs. 3 and 4 in the report by Van der Horst. The head circumference was 14⅝ in. (37.2 cm.). The vault of the scalp was shiny, smooth and uniformly globular, and of a bluish-grey colour. There was no frank fluctuation, but pitting edema, particularly over the forehead and temple region, was very obvious. The fontanelles could not be clearly distinguished, for the whole scalp was tense. It was thought at the time that the fontanelle tension was raised. There were no localizing signs of neurological damage. Because of the edema of the eyelids the ocular fundi were seen only with difficulty after dilatation of the pupils. The retinae appeared normal. The hemoglobin was 6.6 g. %. The red cell morphology was normal for a newborn. The white blood count was 19,000 per c.mm. with 71% lymphocytes. The platelets appeared normal in number. The clotting time was indefinitely prolonged. The prothrombin time (micro method) showed no clot at 120 seconds. The baby's blood was group A, Rhesus positive (D positive). The direct Coombs test was negative.

That the baby had bled copiously was certain, and it was considered that perhaps 175 c.c. might have been lost from the circulation. While clearly there had been some bleeding into the scalp and some blood loss in the skin bruises, at that time it scarcely seemed likely that so much blood could be present at these sites. There was no history of any gastrointestinal hemorrhage, nor any evidence of hemorrhage into the abdomen, such as might occur with a ruptured liver or spleen. It was thought that there must be considerable bleeding within the cranium, unless perhaps the baby had been born exsanguinated from hemorrhage into the maternal circulation. The history did not suggest the latter possibility because the pallor and collapse had occurred after birth.

Whatever the site of the bleeding, the indefinitely long clotting time and the prolonged prothrombin time indicated that the baby had hemorrhagic disease of the newborn. Immediate transfusion was obviously required. Group-compatible and cross-matched citrated blood from the blood bank was given via the umbilical vein. Fifty c.c. was given rapidly during 10 minutes, and a further 80 c.c. was given over the next 3½ hours. Two mg. of vitamin K_1 oxide was given intramuscularly. The baby's condition rapidly improved. Twelve hours later the hemoglobin was 15.8 g. %, and the clotting time was normal, as was the prothrombin time. Whereas previously it had been thought unwise to perform subdural and spinal taps, these were now done. The spinal fluid was lightly blood-stained. No subdural effusions were found.

At first the baby was somewhat lethargic but her general condition improved rapidly. The day after admission the head circumference had diminished to 14¼ in. (36.2 cm.) and soon reached a constant circumference of 13¾ in. (34.9 cm.). After three or four days, orbital hematomata appeared. At no time was the baby jaundiced. At the end of a week she appeared to be behaving nearly normally, but there was a certain tremulous character to her movements. One month later, no definite abnormality could be discovered.

CASE 2.—While the above report was in preparation, a further case was seen. Baby "L", with a birth weight of 9 lb. 1 oz., was born to a primipara after a normal pregnancy and an easy low forceps delivery. No vitamin K was given to either mother or baby.

On the day following the birth, large bilateral parietal cephalhematomata were noted (Fig. 3). Two days later the baby was obviously jaundiced (serum bilirubin 9.8 mg. %) and pale (hemoglobin 10 g. %). The direct Coombs test was negative.

Five days after birth the baby was referred to this institution from the St. Thomas-Elgin Hospital. By that time the infant was very jaundiced (serum bilirubin 15 mg. %) and pale (hemoglobin 8.3 g. %), had a heart rate of 180 per minute, and was quite lethargic. Clotting mechanism studies were not done. A transfusion of 100 c.c. of one-day-old citrated blood was given. The following day the hemoglobin was 12.1 g. % and the serum bilirubin 17.2 mg. %. Although the hemoglobin was rather low, no further transfusion was given.

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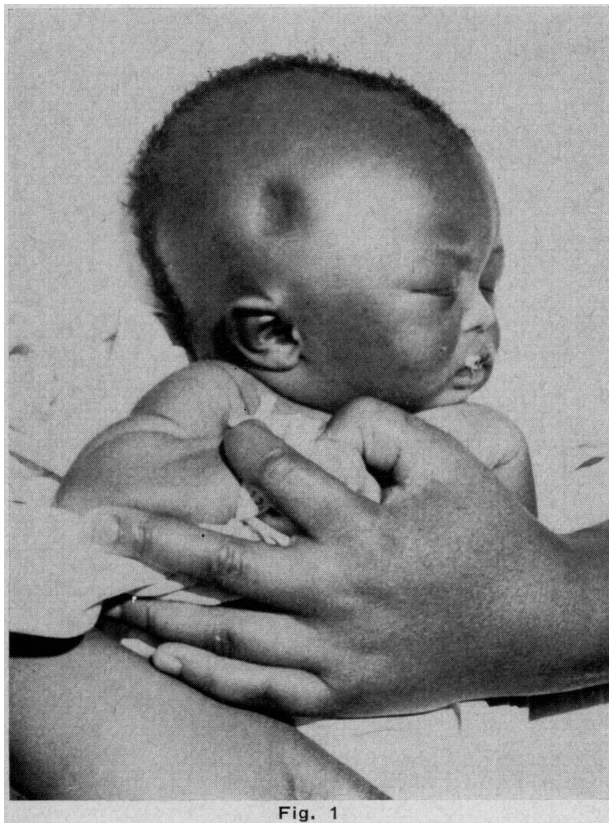

Fig. 1

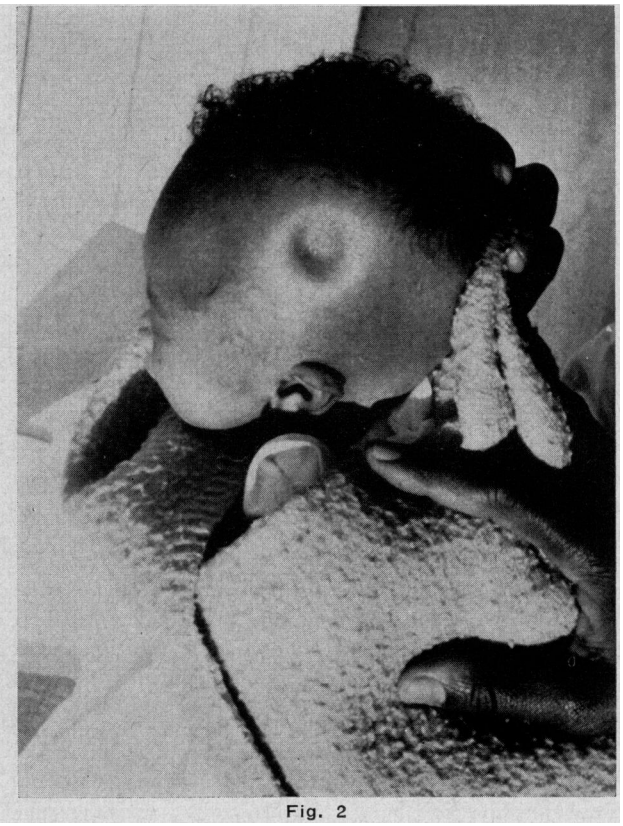

Fig. 2

Figs. 1 and 2.—Exsanguinating subaponeurotic cephalhematomata in two African newborns (Van der Horst's cases). Note uniform enlargement of head, pitting edema and edema of eyelids. (Reproduced with permission from Dr. R. L. Van der Horst and *Archives of Disease in Childhood.*[1])

The infant's general condition was much improved, the jaundice faded quickly and the baby returned home to breast feeding.

The very large bilateral sub-periosteal hematomata had diminished greatly in size by the age of three weeks, but it is anticipated that it will be some months before the skull contour returns to normal.

DISCUSSION

Case 1 is undoubtedly an example of an exsanguinating subaponeurotic cephalhematoma. According to Van der Horst, this lesion is extremely rare in European infants, and this must indeed be so. No mention is made of such a condition in standard pediatric texts or in texts on pediatric pathology. In such writings, the term cephalhematoma is used to refer to the common type of subperiosteal bleeding in which the limits of the hematoma are sharply defined by the sutures of the skull bones, and the volume of blood lost is usually quite small. The subaponeurotic hematoma presents quite differently, for the blood tracks widely over the vault of the skull from the occiput to the nose, and in so doing produces a general enlargement of the head, which is rather deceptive. Because of the uniformity of the cephalic enlargement, one has difficulty in appreciating how great a quantity of blood has been lost from the circulation; yet in the baby described in this report more than half of the normal circulating blood volume had escaped into the subaponeurotic space.

It is strange that in this case (Case 1) no jaundice was evident. One would think that breakdown of such a large volume of blood would certainly give rise to jaundice in the newborn. In Van der Horst's series, there appeared to be no relation between the size of the hematomata as judged by the hemoglobin level and the maximum serum bilirubin level recorded. Indeed, the baby with the second largest blood loss had the second lowest serum bilirubin level. One must evidently be sceptical of the concept that breakdown of blood shed within the body can be a cause of severe jaundice in a newborn.

The baby described in this report (Case 1), like those of the Durban series who were tested, had an indefinitely prolonged prothrombin time. Since this is the diagnostic feature of the condition known as hemorrhagic disease of the newborn, Van der Horst's patients and ours may be said to have had that disorder. However, it is widely accepted that administration of vitamin K to the newborn will prevent hemorrhagic disease of the newborn; and yet four of Van der Horst's 10 cases had been given 2 mg. of vitamin K at birth, a dose which is believed to be effective in prevention of that disease. Since hemorrhagic disease of the newborn is not rare in European babies, and subaponeurotic cephalhematoma seems to be quite uncommon, one wonders what other factors may be involved. Direct trauma to the head does not seem to be causally

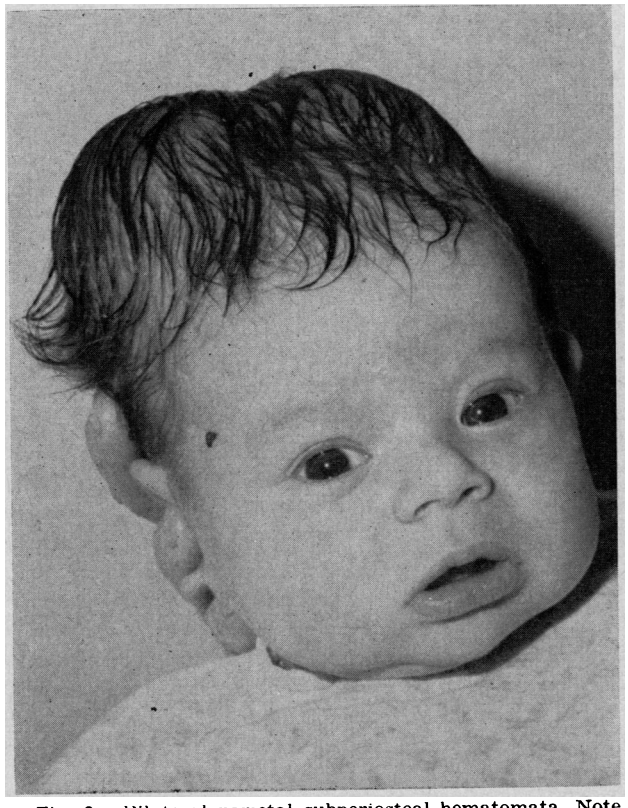

Fig. 3.—Bilateral parietal subperiosteal hematomata. Note localization of the hematoma with limitations set by the cranial sutures.

related, for in three cases of the South African series, delivery was by Cesarean section performed in two instances because a previous section had been done. In our case and in Van der Horst's remaining seven cases, the delivery was not unusual. Van der Horst[2] has the impression that this disorder has a seasonal incidence and considers that a nutritional deficiency may possibly be involved.

Case 2 is an example of the much more usual subperiosteal cephalhematoma. It is not rare for these lesions to be bilateral, but it is decidedly uncommon for sufficient blood to be lost that transfusion is required. While the life of our Case 2 was never in danger, it is clear that the subperiosteal cephalhematoma could kill by exsanguination. Van der Horst's Case 10, an example of subperiosteal hematoma, had a hemoglobin of 3.8 g. %.

Where urgent transfusion of the exsanguinated newborn is required, the umbilical vein provides convenient access to the circulation, which will be found much simpler to use than the small veins of the neonate. It is easily seen as a wide-mouthed, thin-walled vessel in the upper part of the umbilical stump when the cord is sheared off close to the skin. By contrast, the two umbilical arteries are small, tightly constricted, nodular vessels, with no visible lumen. One must remember, however, that in using the umbilicus for transfusion, the circulation is being entered through an area that is colonized very early by potentially pathogenic organisms. A foreign body such as a transfusion cannula should not be left in this position for longer than is necessary. The umbilical vein, because of the dangers of septicemia and portal vein thrombosis, is not a vessel to be used for long periods of infusion.

While donated blood will add prothrombin and Factor VII, and thus of itself cure the coagulation defect, it would seem wise, even if transfusion is given, to give vitamin K_1 oxide also. Whether transfusion is given or not, this appears to be the preparation of choice in the treatment of hemorrhagic disease of the newborn, for its action is more rapid, and the danger of jaundice less, than if some other synthetic analogues of vitamin K are used.

Transfusion for hemorrhage in the newborn can be a matter of the greatest possible urgency. In a calamitous situation, one should not await the cross-matching of blood. Transfusion reactions are extremely unlikely to occur in the newborn, and in a desperate situation the best blood to use is that which can be most rapidly obtained.

SUMMARY

Two cases of exsanguinating cephalhematoma are described. In the treatment of this disorder transfusion may be urgently required as a life-saving measure.

My thanks are due to Dr. E. M. Sharpe, Strathroy, and Dr. C. A. Bell, Port Stanley, Ontario, who kindly referred these patients. Dr. Van der Horst kindly supplied Figs. 1 and 2, which are reproduced by permission of the editors of the *Archives of Disease in Childhood.*

REFERENCES

1. VAN DER HORST, R. L.: *Arch. Dis. Child.*, **38**: 280, 1963.
2. *Idem*: Personal communication.

PAGES OUT OF THE PAST: FROM THE JOURNAL OF FIFTY YEARS AGO

THE THOUGHT OF RARE THINGS

The assembling of possibilities and excluding one after another has all the delights of an intellectual game. Sometimes we are saved from error by our lack of knowledge of the finer points of the game. I well remember a fellow house-officer and myself being much interested in the diagnosis of an obscure abdominal condition. We went over it from every side and to the best of our ability, coming at last to a diagnosis. The attending physician made a diagnosis which had never even occurred to us to consider. He suggested a rare condition which neither of us had ever seen but we felt that consideration of it should not have escaped us. We were in a very humble frame of mind until the operation showed that our diagnosis had been right. It was so principally because the rare condition had not come to our minds. The moral of this is not that ignorance is an advantage. But some of us are too much attracted by the thought of rare things and forget the law of averages in diagnosis. There is a man who is very proud of having diagnosed a rare abdominal disease on several occasions. But as for some years he made this diagnosis in every obscure abdominal condition, of course being nearly always wrong, one cannot feel that he deserves much credit.—T. McCrae, *Canad. Med. Ass. J.*, 4: 587, 1914.