

## CONGENITAL THROMBOCYTOPENIC PURPURA

BY

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Congenital thrombocytopenic purpura is a rare condition. A review of the literature fails to show any case reported from this country. In 1925 Rushmore was able to find seven examples of purpura in pregnancy in which both mother and child were affected, but the type of purpura was not specified. Since that time congenital thrombocytopenic purpura has been recorded on fourteen occasions with eight thrombocytopenic mothers (see table 1). The following instance in a newborn infant whose mother did not suffer from the disease may therefore be of interest.

### Clinical details

A male infant, three days old, was admitted to the Hospital for Sick Children, Great Ormond Street, on April 21, 1944, with a purpuric eruption. He was born at term following an uneventful pregnancy and labour. On the second day of life purpuric spots were first noticed on the buttocks and extended rapidly to the legs and scalp. Jaundice also appeared at this time. His parents and one older sister were healthy, but the second sister suffered from recurrent epistaxis.

**Examination** showed a normally responsive, well-nourished baby, weighing 5 lb. 14 oz. T. 100.2° F. (rectal), P. 140, R. 40. A diffuse purpuric rash was present over all areas of the skin and also on the hard palate with small ecchymoses on the back and legs; mild jaundice was present; mucous membranes were a good colour; the spleen was not palpable; and there was no abnormality of heart, lungs, abdomen or central nervous system. No evidence of infection was found. The urine and stools were free from blood. Pathological investigations carried out on the fourth day were as follows:—

#### INFANT:

Blood count:	R.B.C.	6,700,000 per c.mm.;	Hb.
		130 per cent. (Sahli);	C.I. 0.97.
	W.B.C.	13,300 per c.mm.	
	Polymorph.	58 per cent. with 38 per cent. stab forms.	
	Lymphocytes	37 per cent.	
	Monocytes	1 per cent.	
	Eosinophils	4 per cent.	
	No immature cells seen.		
Platelet count	.. ..	6000 per c.mm.	
Bleeding time	.. ..	10 minutes (Duke's method—normal 2–5 minutes).	
Prothrombin time	.. ..	26 seconds (McTookey's method—normal 11–15 seconds).	
Blood group O	.. ..	Rh-positive.	

#### MOTHER:

Platelet count	.. ..	350,000 per c.mm.
Blood Wassermann reaction		negative.
Blood group O	.. ..	Rh-positive.
No abnormal antibodies were present in the serum.		

The infant remained in hospital for four weeks, during which time he made good progress, gaining 20 oz. in weight. The purpura faded gradually; a few new lesions appeared during the first fortnight; the stools contained streaks of bright blood from time to time and there was one small haematemesis. He was symptom free by the end of the third week. Apart from a temperature of 100° F. on admission he was afebrile, his rectal temperature varying between 97.6° F. and 99° F. for the first eight days and then becoming stabilized at 98° F. No infection was detected at any time. The jaundice continued for eighteen days which is considerably longer than usual in physiological jaundice; the fall in the red blood count was also marked (see table 2). In spite of clinical improvement the thrombocytopenia persisted with a platelet count of only 10,000 per c.mm. on discharge from hospital at one month old (see table 2). The normal figures for the neonatal period are 200,000–300,000 per c.mm. (Sanford, 1942).

The patient was not seen again until he was six months old because of the danger from flying-bombs in the London area. During the interval he had remained symptom free although he had had a severe attack of bronchitis. The platelets had risen to 350,000 per c.mm. and the bleeding time had fallen to 5½ minutes.

This was a spontaneous recovery. The only treatment the child received was vitamin K (5 mgm. kapilon intramuscularly) on admission and liver (Armour's Proethron forte 2 c.cm.) given on the twenty-first and twenty-third days, the latter being without effect on the thrombocytes. Owing to the benign clinical course blood transfusion was not considered necessary.

### Comment

This appears to be an undoubted example of congenital thrombocytopenic purpura. The rash was noticed by the second day and the platelets were reduced to 6,000 per c.mm. on the fourth day. A symptomatic purpura was unlikely as no infection was found, no drug liable to cause a thrombocytopenia had been given to mother or child, and in spite of the prolonged jaundice no other blood disease was demonstrable.

In nine of the cases reviewed the condition was

TABLE 1.—CASES OF CONGENITAL THROMBOCYTOPENIC PURPURA

Reference	Manifestations	Platelet count per c.mm.	Bleeding time (minutes)	Treatment	Course	Family
WALTNER (1924)	Purpura of skin at birth.	90,000	12	None.	Marked icterus; some sepsis. Symptom free by seven months.	Mother: purpura at seventh month of pregnancy. Platelets 50,000. Bleeding time 27 minutes.
LESCHKE (1926)	Purpura of skin at birth with haematuria and melaena.	0	12-15	None.	No bleeding after tenth day.	Mother: purpura at fifth month of pregnancy. Platelets 60,000.
LIEBLING (1926)	Purpura of skin at birth with haematemesis and melaena.	40,000	—	Haemoplastin, 5 c.cm. intramuscularly.	Did well. Purpura faded at one month. Platelets 240,000.	Mother: purpura at fifth month of pregnancy. Platelets 40,000.
GREENWALD (1929)	Purpura of skin appeared on fifth day.	30,000	5	None.	Died on seventh day. Congenital morbus cordis. Thrombocytes scanty and abnormal in appearance.	—
BAYER (1931)	Purpura of skin at birth.	50,000	40	None.	No fresh bleeding after birth. Platelets 320,000 by second year.	—
Ibid.	Purpura of skin at birth.	48,000	6½	None.	No fresh bleeding after birth. Platelets 150,000 on fifth day.	Twin. Platelets 77,000. Bleeding time 1 minute. No symptoms.
Ibid.	Purpura of skin and mucous membranes at birth with haematemesis and melaena.	45,000	11	Coagulen and gelatin intramuscularly.	No fresh bleeding after fifth day. Platelets 280,000 by second year.	—
Ibid.	Purpura of skin at birth. Haematemesis on second day.	50,000	90	None.	Died on second day. Widespread purpuric lesions.	—
GUTFREUND (1933)	Purpura of skin and mucous membranes at birth.	0	40	90 c.cm. blood intramuscularly, also blood transfusion (? amount).	Continued to bleed. Died of pneumonia at four months.	Father: severe epistaxis and excessive bruising. Mother: Platelets 32,000, but symptom free.
SANFORD (1936)	Purpura of skin at birth.	20,000	—	None.	No fresh lesions after birth. Platelets 137,000 by tenth day. Mother and child both showed increased disintegrating power of platelets.	Mother: recurrent purpura. Platelets 17,500.
DAVIDSON (1937)	Purpura of skin at birth.	56,000	3	Blood, 100 c.cm. intramuscularly without improvement. Bothrops antitoxin, 5 c.cm. intramuscularly with clinical recovery.	Platelets 672,000 by ten weeks. Remained normal for two years.	Mother: splenectomy eight years before for thrombocytopenic purpura. Platelets 6,000-50,000.
WHITNEY (1942)	Purpura of skin at birth.	80,000	1	Blood intramuscularly, 20 c.cm.	Died third day. Tentorial tear. Widespread purpura.	Mother of these two infants had splenectomy at fourteen years of age. Platelets 70,000.
Ibid.	Purpura of skin at birth.	90,000	11	Blood intramuscularly, 55 c.cm., twice.	Died second day with multiple haemorrhages.	
URBANSKY (1942)	Purpura of skin and mucous membranes.	23,200	5½	50 c.cm. blood intravenously.	Purpura faded in a few days. Bleeding time 1½ minutes at 2½ years old.	Mother had splenectomy six years previously for thrombocytopenic purpura. Platelets 75,000 at six months of pregnancy.

TABLE 2

Date	Hb. per cent.	R.B.C. per c.mm.	W.B.C. per c.mm.	Platelets per c.mm.	Bleeding time
22.4.1944	130	6,700,000	13,300	6,000	10 minutes +
1.5.1944	80	3,500,000	22,200	7,000	—
8.5.1944	80	4,100,000	16,000	15,000	10 minutes +
12.5.1944	—	—	—	12,000	—
26.5.1944	—	—	—	10,000	—
24.10.1944	—	—	—	350,000	5½ minutes

familial; in the four collected by Bayer the mothers' platelet counts were not noted. In the present instance the mother's blood was normal and she had never shown a tendency to bleed.

The prognosis in these infants is of some interest. Nine of the fourteen recovered during the first few weeks of life but their progress was not followed further except in three who remained healthy for two years. It would be instructive to know whether or not these children tend to relapse later in life. Five of the infants died, associated lesions may have been the cause of death in three of them. Two only died of uncomplicated purpura. Thus prognosis does not appear to be so grave as might be expected.

From a study of these reports therapy seems to have had little effect. Five infants recovered without treatment. Blood transfusions in adequate amounts would seem to be the rational treatment in any severe case. The advisability of splenectomy at birth was discussed by Whitney (1942) but in view of the above figures does not appear justifiable.

#### Summary

A case of congenital thrombocytopenic purpura is reported.

Examples of congenital thrombocytopenic purpura collected from the literature are tabulated.

The prognosis and treatment of the condition is considered.

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