ACUTE BUDD-CHIARI SYNDROME*

Report of a Case

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The Budd-Chiari syndrome is extremely rare in children and uncommon in adults. About 170 cases have so far been reported in the literature. The diagnosis is often incidental or accidental at the time of post-mortem examination. This case is being reported because of its rarity in clinical practice and the fact that the child is still alive.

Report of a Case

A female child aged 7 years was admitted with acute abdominal pain of 5 days’ duration. The pain was sudden in onset, excruciating in nature, and did not radiate. It was continuous and did not vary in intensity. The child used to adopt a peculiar sitting posture i.e. would bend forward supporting herself on her arms extended out in front. Vomiting was a constant feature at the onset. The vomitus contained bile-stained material besides food ingested previously. It was not foul smelling and did not contain any blood. There was no history of fever or cough. Stools were normal. There was no urinary complaint. The past and family history was non-contributory. The child had not taken any drug in the preceding two weeks, was non-vegetarian and had been in good health all along.

Examination showed a moderately nourished, slightly anaemic child who was ill-looking. Pitting edema of the feet was present and extended up to the knees. There was no jaundice, cyanosis or clubbing. The tongue was moist. The neck veins were not engorged. The hepato-jugular reflex was absent. Mild congestion of the throat was found. A small lymph gland was palpable in the right axilla. The temperature was normal, pulse 120/min. and B.P. 120/80 mm of Hg.

The abdomen looked full and it moved well with respiration. No engorged veins were seen over the abdominal wall. The umbilicus was normal in position. There was no rigidity. The liver was palpable 3 cm. below the costal margin. It was tender, with a smooth surface and firm consistency. The spleen was just palpable. There was free fluid in the peritoneal cavity. Peristaltic sounds

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were heard normally. The cardio-
vascular system was normal except
for tachycardia. The rest of the sys-

temic examination was normal.

Laboratory investigations. Hemoglo-
bin was 7 Gm. %, total white cell count
25,000/cmm, polymorps 63 %, eosino-
phils 10 % and lymphocytes 27 %. Bleed-
ing time was 2 min. 20 seconds,
clotting time 2 min., prothrombin
time 25 seconds, blood urea 107 mg %,
van den Berg test negative, Gross test
positive, Hanger test ++, Maclagan
test 4.4 units, serum bilirubin 1 mg %,
serum alkaline phosphatase 8 units,
total serum cholesterol 125 mg %, serum
amylase 124 units and blood sugar
100 mg%. The stools showed Enta-
moeba histolytica (trophozoite form).
A liver biopsy revealed hypopro-
teinemic changes. Specific gravity of the
urine was 1013, acidic, albumin+, with
a large number of pus cells and a few
red blood cells. Urobilinogen was ++
and there was no sugar. Urine culture
was sterile.

Treatment. The child received pethi-
dine, intravenous fluids, heparin,
Achromycin, corticosteroids and intra-
venous liver extract. Following this
treatment she developed pulmonary
oedema, which was attributed to
excessive intravenous fluids. Forty-
eight hours later, the child began passing
blood in the faeces. Heparin was
stopped and an antidote given. Anti-
amoebic treatment was started with
Humatin (Parke-Davis) and chloro-
quine. Thereafter, she felt better and
there was little pain. The temperature
came down and there was no tender-
ness over the liver. The lungs were clear
and urine was found to be normal. The
patient had completely recovered from
her illness and liver biopsy was done
one day before discharge. When seen
one month after her illness, she was
found to be perfectly normal.

Discussion

With these physical findings, urinar-
year abnormalities, raised blood urea,
absent hepato-jugular reflex, oedema of
the feet and ascites, a diagnosis of acute
Budd-Chiari syndrome was thought of.
The syndrome classically manifests
itself as hepatomegaly, shock, coma,
ascites, splenic enlargement, oedema
and occasionally jaundice. In the diffe-
rential diagnosis we considered acute
pancreatitis, acute nephritis and acute
amoebic hepatitis but there was no
supportive evidence. In our case, the
features of hepatomegaly, shock,
ascites, splenomegaly and absent hepa-
tojugular reflex can all be explained by
the diagnosis arrived at. Liver biopsy
was not done in the acute stage for
fear of precipitating massive haemo-
rhage. Just before discharge, it was
performed and showed hypopro-
teinemic changes. We feel that the throm-
bosis had been precipitated by infec-
tion, as was also seen in a previous
case (Gupta, 1965).

The etiology of the condition cannot
always be established. Primary vas-
cular disease, spontaneous thrombosis,
congenital malformations of the veins
and pathological processes in the vic-
nity causing pressure on the vessels
may all produce the syndrome (Popper
and Schaffner, 1957). It is known to
occur in association with cirrhosis,
obliterative endophlebitis, tumours,
abscesses, syphilis, veno-occlusive
disease, poisoning with senecio alka-
loids, retrograde embolization and
polycythemia vera.

One must be aware of the condition
to be able to diagnose it during life.