

# Splenectomy in onyalai

## A report on 5 cases

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### Summary

Onyalai is an acquired immune thrombocytopenia with 10% mortality. Conservative measures such as traditional medicines, corticosteroids and blood transfusion have not always controlled severe bleeding or prevented death. Five patients (2 male, 3 female) with onyalai who had uncontrollable haemorrhage, thrombocytopenia and documented previous attacks of severe bleeding, underwent splenectomy. The patients were screened for malaria, sickle-cell anaemia and bilharzia. Vitamin K and  $\epsilon$ -aminocaproic acid were administered pre-operatively, and fresh blood was given during surgery. The duration of follow-up varied between 280 and 544 days. There were no operative complications. Bleeding stopped in all patients and the platelet counts increased within 24 hours. All achieved normal platelet counts, but these were not always sustained. Three patients remained free of disease with normal platelet counts up to days 539, 539 and 544 of follow-up. Two patients had a recurrence of bleeding and died from cerebral haemorrhage and haemorrhagic shock.

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Onyalai, a condition described by Wellman in 1904, is an acquired bleeding disorder mainly limited to certain negroid populations in Africa. The incidence of onyalai in the Kavango territory of SWA/Namibia was 1:660 in 1981.<sup>1</sup> The laboratory features include profound thrombocytopenia with abundant megakaryocytes in the bone marrow,<sup>2</sup> and the presence of IgG and IgM platelet antibodies in the serum.<sup>3</sup> Previously healthy people suddenly become ill; haemorrhagic bullae appear in the buccal mucosa and the skin and severe haemorrhage — from which 10% of patients may die through blood loss or cerebral haemorrhage — follows. The natural course is usually one of prolonged asymptomatic periods of thrombocytopenia, interspersed with episodes of clinical bleeding.<sup>1</sup> The aetiology is unknown but, although idiopathic thrombocytopenic purpura (ITP) and onyalai have many features in common, there is enough difference to consider onyalai a separate disease entity.<sup>1</sup>

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The optimal management of onyalai has not yet been determined. A review of the literature reveals that among other things various indigenous plants,<sup>4</sup> cod-liver oil,<sup>5</sup> arsenic and lead, quinine, ergot, vitamins, thromboplastin-like substances,<sup>6</sup> sex hormones and intramuscular injections with blood<sup>7</sup> have been tried without real success. In some patients the replacement of lost blood and the administration of fresh blood does not prevent cerebral haemorrhage.<sup>1</sup> Because of the similarities with ITP, certain investigators have used prednisolone in the treatment of onyalai with equivocal results.<sup>3,8-11</sup> The case histories of 2 patients with onyalai who underwent splenectomy and had normal platelet counts 6 weeks later have been recorded.<sup>9</sup>

In view of the high mortality rate in the acute phase of onyalai, 5 patients at high risk of fatal haemorrhage underwent splenectomy and were followed up to assess the value of this procedure.

### Patients and methods

#### Case 1

A 59-year-old man, who had experienced attacks of onyalai in childhood and during the previous 6 years, had been bleeding continuously from onyalai for 3 months. On examination he had numerous haemorrhagic bullae in the buccal cavity and the skin. The platelet count was  $8 \times 10^9/l$  and the haemoglobin value was 7,7 g/dl. A splenectomy was performed 4 days after the patient's admission to hospital.

#### Case 2

A 38-year-old woman required numerous blood transfusions during attacks of onyalai in 1980 and 1981. Clinical disease recurred a year later with severe blood loss requiring three blood transfusions. Examination revealed numerous haemorrhagic bullae in the buccal cavity and the skin. The platelet count was  $3 \times 10^9/l$  and the haemoglobin value was 12,5 g/dl. A splenectomy was subsequently performed.

#### Case 3

A 38-year-old woman experienced severe attacks of onyalai in 1980 and 1981. Another attack occurred during the following year when she was 8 months' pregnant. When she went into labour there were actively bleeding haemorrhagic bullae in the buccal cavity. The next day her platelet count was  $11 \times 10^9/l$ . Postpartum haemorrhage persisted despite the administration of 3 units of fresh blood. A splenectomy was performed a week later in an attempt to stop the bleeding. At birth the infant showed no clinical signs of a tendency to bleed and had a platelet count of  $210 \times 10^9/l$ .

#### Case 4

A 14-year-old boy had experienced between one and three attacks of onyalai annually since his 8th year, these necessitating numerous blood transfusions. The last attack occurred in March 1982. No physical signs of onyalai were present on clinical examination a year later. His platelet count at that time was  $59 \times$



$10^9/l$  and his haemoglobin value was 11,1 g/dl. A splenectomy was performed.

### Case 5

A 30-year-old woman had had an attack of onyalai as a child and a well-documented attack in May 1981 which was accompanied by severe haemorrhage. The condition recurred 2 years later. She received several blood transfusions over the next month, but the haemorrhagic bullae and bleeding persisted until her spleen was removed. The pre-operative platelet count was  $26 \times 10^9/l$ .

All these patients conformed to the criteria of Wiseman *et al.*<sup>11</sup> for the diagnosis of ITP. The following procedures were carried out on each patient pre-operatively: full blood count, bone marrow aspiration, liver function test, electrolyte and urea measurements, sickling test, malaria smear test, prothrombin index determination, urinalysis, stool microscopy and chest radiography.

Ten milligrams of vitamin K<sub>1</sub> was administered 24 hours pre-operatively and 8-hourly  $\epsilon$ -aminocaproic acid was commenced 24 hours before the operation. An infusion of fresh blood was started as surgery began. The anaesthetic agents used for all the patients were sodium pentothal, alcuronium chloride, halothane, nitrous oxide and oxygen. The splenectomies were performed via a left abdominal paramedian incision.

The patients were followed up at regular intervals for a period of 280-544 days after the splenectomy; at each visit a clinical examination and full blood count were carried out.

### Results

The operation was well tolerated by all the patients and no intra- or postoperative complications occurred. The total blood loss during the operation varied from 100 ml to 500 ml. Diffuse bleeding from all the cut surfaces diminished markedly soon after the pedicle of the spleen had been clamped. Patient 1 had a haemorrhage into the visceral peritoneum. Patient 2 had no visible evidence of a haemorrhagic tendency in the abdominal cavity. Patient 3 had numerous splenic infarcts and numerous petechiae in the omentum, parietal peritoneum, surface of the uterus and the fallopian tubes. Patient 4 had no visible signs of a bleeding tendency in the abdominal cavity. Patient 5 had numerous petechiae throughout the abdominal cavity.

The results of the pre-operative investigations are listed in Table I. The changes in the platelet count during the first 14 days after splenectomy and during follow-up are illustrated in Fig. 1. The platelet count more than doubled in every patient within 24 hours of the operation. The platelet count subsequently reached normal levels in all the patients.

Patient 1 suffered a clinical recurrence of onyalai on day 319 of follow-up and died of cerebral haemorrhage on day 330. Patient 5 presented with a clinical recurrence of onyalai on day 280. She was referred to Rundu hospital, but died *en route* from haemorrhagic shock. The other 3 patients have maintained adequate platelet counts and remained in good health during the follow-up period of 539 - 544 days. Malaria smears were negative in all patients at their follow-up visits.

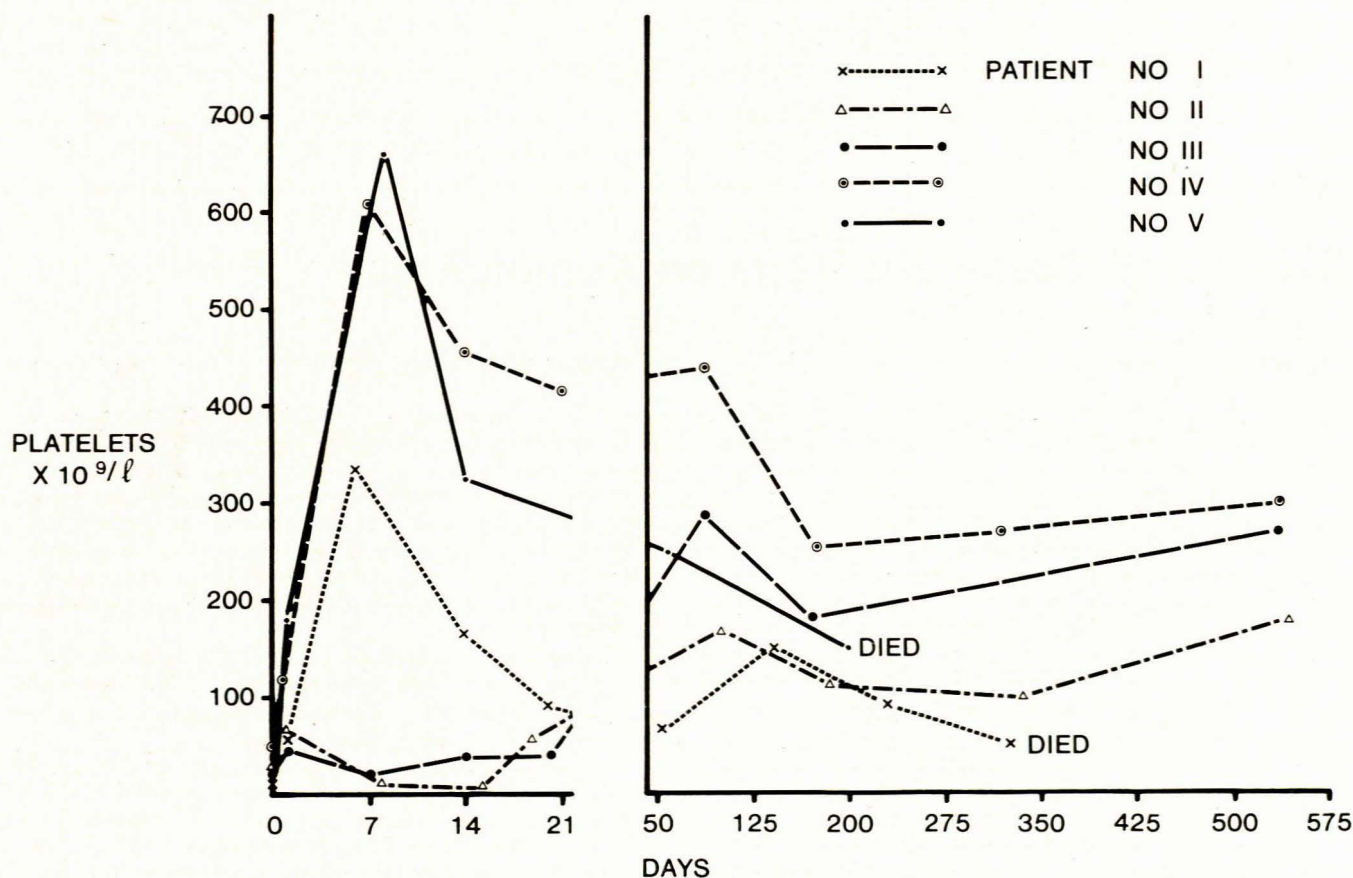


Fig. 1. Changes in the platelet count after splenectomy.



**TABLE I. RESULTS OF PRE-OPERATIVE INVESTIGATIONS IN 5 PATIENTS WITH ONYALAI**

	Case				
	1	2	3	4	5
Haemoglobin (g/dl)	7,7	12,5	9,9	11,1	13,2
White cells (x 10 <sup>9</sup> /l)	3,5	7,7	10,1	12,0	5,6
Platelets (x 10 <sup>9</sup> /l)	8	3	16	59	26
Electrolytes and urea	N	N	N	N	N
Liver function test	N	N	N	N	N
Prothrombin index (%)	90	94	100	94	95
Chest radiography	N	N	N	N	N
Malaria smear	Neg	Neg	Neg	Neg	Neg
Sickling test	Neg	Neg	Neg	Neg	Neg
Stool microscopy	Rbc	N	Rbc	N	N
Urinalysis	Rbc	Rbc	Rbc	N	N
Bone marrow aspiration	M	M	M	M	M

N = normal; Neg = negative; Rbc = red blood corpuscles; M = normal with a normal or increased number of megakaryocytes.

## Discussion

Splenectomy was followed by a rise in the platelet count and cessation of bleeding in all patients. No operative or postoperative complications occurred and blood loss was within normal limits. Two patients experienced a clinical recurrence of onyalai. One died of cerebral haemorrhage and the other of haemorrhagic shock. Three patients are alive and well.

We conclude that splenectomy is a useful measure to raise the platelet count and arrest bleeding in patients who do not respond to conservative treatment, but are of opinion that it is indicated in onyalai only when bleeding cannot be controlled or if signs of central nervous system haemorrhage develop, when the operation should be performed as an emergency procedure.

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# News and Comment/Nuus en Kommentaar

## Wind and water

Players of wind instruments have problems of which players of other instruments are blissfully unaware. Among these is the accumulation of saliva in the instrument which, if it accumulates in the wrong place, can metamorphose a soulful rendition of a melody into a distinctly unmusical, agricultural-sounding croak. A transdermal preparation of scopolamine (for the prophylaxis of motion sickness) is now available, and since it dries the mouth, it was administered in an attempt to improve the quality of note production in players plagued by this threat to their virtuosity (Dettman, *N Engl J Med* 1984; **310**: 1396). The treatment appears to work well. The transdermal patch takes effect 15 minutes after it is administered, and the effect is maintained until about half an hour after its removal. No other side-effects have been noted, although it is worth remembering that scopolamine has been used as a central nervous system sedative, when it was dubbed 'twilight sleep'. This could be disconcerting to a player who has a lengthy rest before the next musical passage. Such a possibility brings to mind the story of the violinist who dreamed that he was playing second violin in Handel's *Messiah* (notorious for its demands in violinists) and woke up to find that he was.

## Contraceptive sponges

A number of governments, including those of the USA and the UK, have recently approved the use of a disposable, over-the-counter contraceptive sponge. This sponge is made of polyurethane and is designed to be compatible with the vaginal environment. During manufacture a spermicide is incorporated into the sponge, which also contains preservatives. It is round, about 5,5 cm in diameter and 2,5 cm thick, and is available in a single size. A polyester loop attached to the sponge makes removal easy.

Effectiveness is due primarily to the release of spermicide into the vagina and not to the barrier function. Clinical trials in a number of countries have suggested that it is reasonably effective but not as effective as the diaphragm.

There are several advantages of the sponge: it can be inserted at least 24 hours before intercourse, it does not have to be fitted, it will give protection against pregnancy for at least 24 hours regardless of the amount of intercourse and, of course, it can be purchased without a prescription and is easy to use. Use of the sponge is reviewed by D. A. Edelman in the June 1984 *IPPF Medical Bulletin*.