

## A CASE OF ANO-RECTAL AGENESIS: POSTOPERATIVE COMPLICATIONS

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### Summary:

**An analysis of the postoperative complications of a case of ano-rectal agenesis is here presented. The multiplicity of factors involved, with special emphasis on protein-calorie malnutrition, together with the importance of team-work in the management of this case, is emphasised.**

### Case Report:

The child was born at St. Luke's Hospital on 6.11.69 of a para-3 mother, after a normal pregnancy and delivery at term. The birth weight was 3.6 Kg. (8 lbs.). At birth he was found to have 'imperforate anus' of the ano-rectal agenesis type and meconium was seen coming out of the urethra indicating the presence of a fistula. The next day a transverse colostomy was performed (R.A.). The following day he developed generalised twitchings which were easily controlled with paraldehyde and he was put on 'prophylactic' antibiotics. His subsequent progress was uneventful and he was discharged home at the age of 7 weeks weighing 4.9 Kg. (10 lb. 10 oz.).

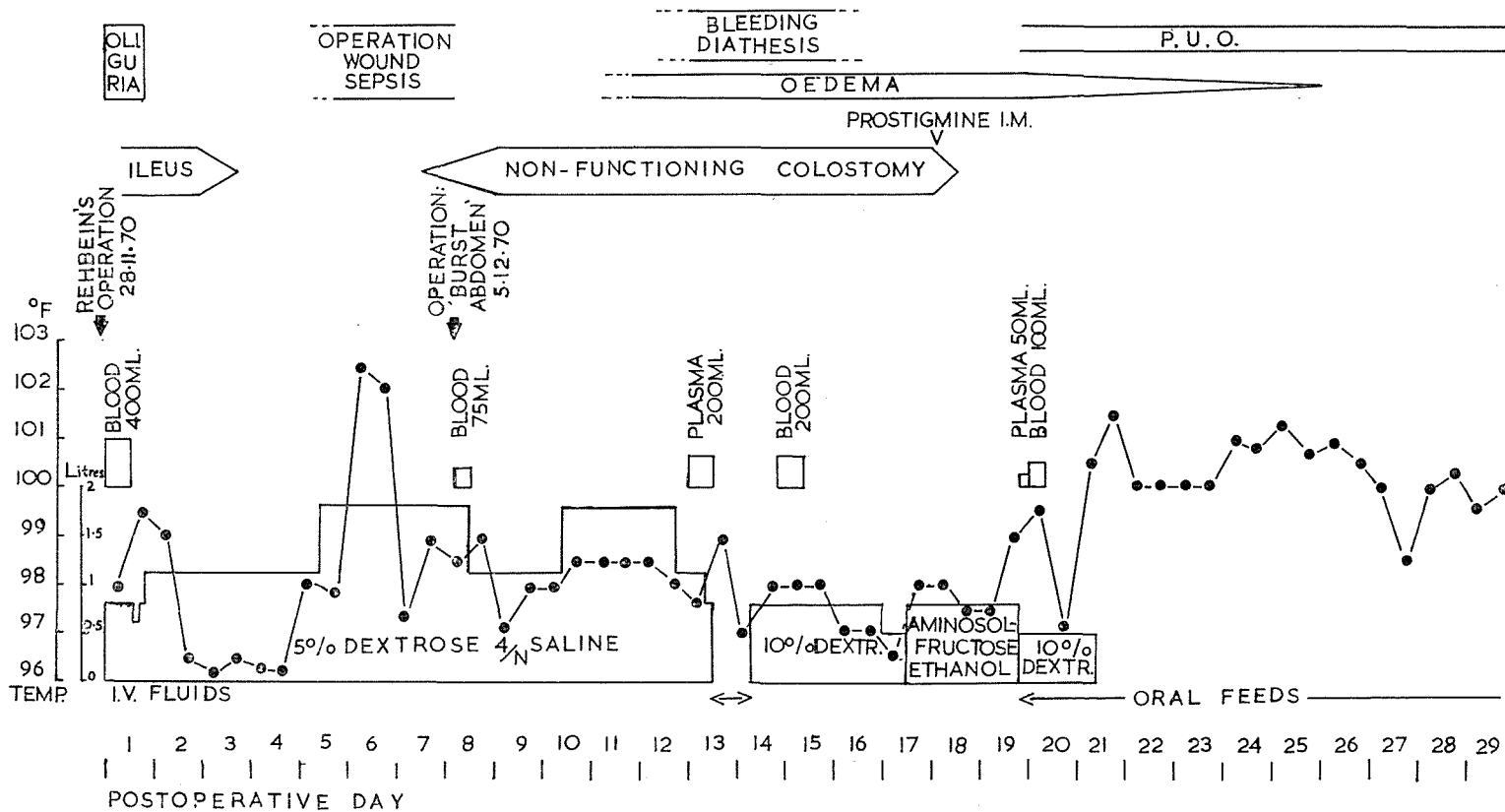
He was admitted to the children's ward on several occasions under the care of Dr. T. Agius Ferrante: at 16 weeks with gastro-enteritis; at 5½ months with severe anaemia (Hb 7.9 G%) and vomiting; at 9 months with severe anaemia (Hb 8.4 G%) and diarrhoea; and at 10½ months with anaemia (Hb 8.8 G%) and chest infection. At 11 months of age (28/10/70) he was admitted to the surgical ward for further surgery on his ano-rectal agenesis, but operation was postponed because of recurrent upper respiratory tract infections.

When 1 year old (28/11/70) and weighing 7.2 Kg. (16 lbs.) the Rehbein pull-through abdomino-perineal operation (Rehbein, 1959) was performed (R.A.) (see fig.). The recto-urethral fistula was tied and the colon, after dissection, was passed through the rectal tube (denuded of its mucosa) and tethered to the perirectal tissues with the end sutured in eversion to the anal margin. The colostomy was not touched at this stage. During and after operation, the child was given 400 mls. of blood and intravenous fluids were continued as 5% dextrose N/4 saline at the rate of 800 mls. per 24 hours. The next day, since he was pyrexial and rales were heard over both lung fields he was started on i.m. Ampicillin and Cloxacillin and the drip rate was reduced to 640 mls. per 24 hours. At this time, the urine output was very poor and a few hours later the infusion rate was increased to 1200 mls. per 24 hours and the antibiotics were continued intravenously. His urine output improved and by the second postoperative day he was afebrile and progressing well. The colostomy worked on the third day, but the abdomen was still distended and the child vomited several times. At this time he was given intravenous potassium supplements (14 mEq./lit of K<sup>+</sup>) and the gastric aspirate was replaced with equivalent amounts of normal saline via the drip. Serum electrolytes and blood urea were monitored as necessary.

On the sixth postoperative day, the patient's condition deteriorated: he became pyrexial (Temp 102.5° F) and had a sinus tachycardia of up to 200/min; his abdomen was very distended, bowel sounds became infrequent and pus could

V.M: ♂ 1yr. ANO-RECTAL AGENESIS

Hb. G% [ % ]	14.8 (100)	13.4 (91)	11.1 (75)	9.7 (66)	8.3 (56)	6.0 (41)
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be seen exuding from the operation wound. He was digitalised and the fever and tachycardia settled over the next 24 hours. An erect X-Ray of the abdomen showed fluid levels and on 5/12/70 (i.e. 8 days after the first operation) laparotomy was performed. When the skin sutures were removed it was obvious that the abdomen had "burst" and the small intestine was distended throughout. This was decompressed by a tube passed via the colostomy and the abdomen was closed by through and through tension sutures. Postoperatively following blood transfusion he was continued on intravenous fluids. By the 8/12/70 the colostomy had still not functioned and the child was noted to be developing generalised oedema, though there was no clinical evidence of circulatory overloading and no albuminuria. Serum electrolytes and blood urea were normal but the total serum proteins (4 G%) and, in particular the albumin (2.1G%), were low.

He was given 200 mls. Plasma and Frusemide (Lasix) 10 mg. (1 mg./Kg.) IM. He was also tried on oral feeds in an attempt to provide calories but he vomited these immediately. By this time, his general state had grossly deteriorated. He was very pale and listless with generalised pitting oedema, distended abdomen, an infected operation wound, poor urine output and a still non-functioning colostomy. He also had numerous ecchymoses and bled easily from venepuncture sites and from the tension sutures. On 12/12/70 he was given more blood, daily i.m. injections of Vitamin K were started and IV fluids were recommended as 10% dextrose. On 14/12/70 because of shortage of suitable peripheral veins, a 'cut-down' was performed on the left long saphenous vein in the groin and a 12 inch nylon IV cannula was inserted via the femoral vein up into the inferior vena cava. Through this he was given a high-calorie solution of Aminosol-Fructose-Ethanol (Aminosol-Vitrum).

Meanwhile, the colostomy had still not functioned 10 days after the second operation and Prostigmine 0.2 mg. IM. was given. About 4 hours after this the

colostomy started functioning. The aminoacid solution was discontinued after 3 days. Afterwards, while the child was being given more plasma, he was reported to have collapsed, becoming pale and limp, with rapid respirations. The plasma was stopped and blood, hydrocortisone and oxygen were given. His general condition slowly improved and by the next day (17/12/70) he was well enough to be started on 10% dextrose by mouth and the IV fluids were reduced in amount. The oedema had by now greatly improved and was limited to the lower limbs and the bleeding diathesis had resolved. On 18/12/70 he was started on milk feeds and solids together with a multivitamin preparation. He then became febrile and had loose stools and was treated with a short course of oral Sulphaguanidine and Streptomycin suspension (Guanimycin). The diarrhoea settled, but the pyrexia persisted for 2 weeks before it resolved spontaneously without chemotherapy. By the 23/12/70 the oedema had virtually disappeared and the main problem was then one of severe anaemia (Hb 6.0 G%). He was treated with intramuscular, and later oral, iron and on the 8/1/71 a repeat Hb was 13 G%.

On 18/12/70 it was noticed that the child could not bear weight on his right leg, keeping it flexed at the hip. X-Ray showed changes suggestive of metaphysitis of the right femur. This however settled within a few days with simple passive movements of the hip joint.

Following this, progress was uneventful and the child underwent two further operations under general anaesthesia. On 18/2/71 the anal orifice was dilated and on 8/3/71 the colostomy was closed. The postoperative periods were both smooth and the child was discharged home well on the 6/4/71 with a Haemoglobin of 13.7 G%. He is to be re-admitted in a few months' time for trimming of the rectal stump.

#### Comments:

##### (1) Postoperative oliguria

This was, in part due to the fact that the child initially received less than the

basal daily fluid requirements for his weight, i.e. 640 to 800 mls. rather than 1200 mls. The blood urea was however never elevated, showing that good renal function was maintained throughout the intra- and post-operative periods. The oliguria was probably largely a manifestation of the normal postoperative response to surgical stress which causes increased antidiuretic hormone and corticosteroid release with water and salt retention. Marked oliguria is nevertheless often a worrying feature in an ill child and a useful test is to increase the rate of infusion over half an hour and to observe its effect on the urine output. This was done in our patient with a brisk diuretic response.

It must also be remembered that in these patients additional fluid and solute are lost via the skin and the respiratory tract, (especially in the presence of pyrexia) and via the gastro-intestinal tract through gastric aspiration, vomiting and colostomy secretions. In the presence of ileus one must also make allowances for water and solute sequestered in loops of dilated bowel. All this requires careful collection and measurement of all losses and meticulous input-output charting on a fluid-balance sheet.

### **(2) Non-functioning Colostomy:**

The colostomy failed to function at all for about 10 days after the second operation, though bowel sounds could be heard on and off on several occasions. This was probably due to a multiplicity of factors among them hypokalaemia, oedema of the bowel wall, wound infection and the poor nutritional status of the child. Serum potassium level was normal but as this is a poor index of the state of the intracellular potassium, the child was nonetheless given K supplements intravenously. Cholinergic drugs to stimulate intestinal peristalsis are rarely indicated because other measures especially gastro-intestinal suction and proper hydration usually suffice. In this patient, the colostomy started working about 4 hours after administration of Prostigmine.

### **(3) Oedema:**

This was not an unexpected complication as the child had been on a grossly inadequate protein-calorie intake during his first 12 postoperative days. A 7.2 Kg. (16 lb.) child in good health would require a basal intake of about 800 calories (100 cal/Kg.) and 24G protein (3G/Kg.) per day. The patient had 5% dextrose N/4 saline as basic intravenous fluids supplying him with 240 to 360 calories per day depending on the amount of fluid intake. The only source of protein was two blood transfusions (given on two separate days) which supplied about 14G protein in all. The oedema was therefore due to protein-calorie malnutrition causing hypoproteinaemia. Moreover, the possibility that the child may have been hypoproteinaemic before operation is unlikely because he had a normal haemoglobin. Plasma was given, but this is only of limited benefit to maintain the intravascular osmotic pressure, thereby slowing the progress of the oedema. Moreover, the calorific value of plasma is only 20 calories per 100 ml. which is clearly inadequate. For these reasons it became obvious that the child needed urgent parenteral nutrition and until the Aminosol-Fructose-Ethanol solution was made available to us we gave 10% dextrose (supplying about 320 calories per day) via a small peripheral vein.

Aminosol contains in the physiological L-form, all the 8 essential and 10 non-essential aminoacids. An absolutely vital factor in the metabolic utilisation of nitrogen is that adequate non-protein calories must be given simultaneously (Michener and Law 1970). These are provided by fructose and ethanol, making up the high-calorie solution (about 875 calories per litre). As this is hypertonic and irritant to vessel walls, a venous catheter was threaded up the inferior vena cava and the solution was given at the rate of 800 mls. per day (100 ml/Kg./day) for three days. Further plasma, blood and 10% dextrose were then given and over the next few days the oedema became less obvious and after a week it virtually disappeared. Once oral feeds are re-established in such cases, rapid improvement usually follows.

**(4) Bleeding Diathesis:**

This was probably due to several factors. Non-availability of protein for synthesis of the various coagulation factors and impaired liver function with failure of synthesis of the Vitamin-K-dependent factor (II, VII, IX, X) were perhaps the two most important mechanisms. It is also likely, that Vitamin K itself may not have been readily available for absorption from the large bowel because the patient had been on broad spectrum antibiotics (Ampicillin and Cloxacillin) for some time before. On 14/12/70 the platelet count was 112,000/cu.mm., so that thrombocytopenia may have been an additional factor. Coagulation studies were not performed. The patient was treated with plasma, blood transfusions and i.m. Vitamin K<sub>1</sub> injections and the bleeding tendency settled after about 5 days.

**(5) Anaemia:**

The child had previously been admitted to hospital with moderately severe anaemia but his haemoglobin levels on the second and fourth postoperative days were normal. The Hb dropped to 11.1 G% around the time when it was obvious that the operation wound had gone septic. The film later on showed microcytosis and hypochromia and the anaemia was most likely due to a combination of infection, iron deficiency and protein-malnutrition. Also, repeated blood sampling by venepuncture can, in a child of this age, amount to quite a considerable degree of blood loss. Using microtechniques, needing as little as 0.1 ml. capillary blood (obtained by the heel-prick method) would avoid this complication.

By the 27/12/70 the child's haemoglobin had fallen to 6.0 G% and therapy was started with intramuscular and later oral iron. The poor initial response to this was probably due to the concomitant presence of infection which is known to interfere with incorporation of iron into the haemoglobin molecule.

**(6) Pyrexia of unknown origin:**

The cause of this fever remained obscure. There was always the great risk that the operation wound might become 'septic' because of difficulty in separating it from the colostomy area. Repeated swabs from the wound site grew mixed flora including *Proteus* and *Pseudomonas* organisms. Blood culture (17/12/70) and urine culture (22/12/70) grew similar organisms, but the significance of these results was doubtful because of the close proximity of the colostomy to the operation wound, to the I.V. cut-down at the groin and to the genital region, in a small child. Although urinary tract infection, or even 'low-grade' septicaemia via the venous catheter seemed likely possibilities at the time, it was decided to withhold antibiotic therapy as apart from his fever the patient was progressing very satisfactorily. The fever finally settled after 14 days.

**Conclusion:**

The postoperative care of a seriously ill child who has undergone major surgery and may be suffering from multiple complications, necessitates close liaison between the surgeon, the paediatrician and the laboratory and nursing staff. It is clear from the comments about this case that the utmost attention to detail is crucial in the management of such cases.

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