Table 1 Neutrophil granule protein concentrations

	Patient	Control
Myeloperoxidase (ng/ml)	9744	21312
Defensins (ng/ml)	0	3808
Human cationic antimicrobial protein (hCAP18) (ng/ml)	87	51200
Lactoferrin (ng/ml)	0	3920
Neutrophil gelatinase associated lipocalin (NGAL) (ng/ml)	7	2575
Gelatinase (ng/ml)	537	7478

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Bilateral thoracoscopic splanchnicectomy in chronic intestinal pseudo-obstruction: report of two paediatric cases

Chronic intestinal pseudo-obstruction (CIPO) is a rare, highly morbid disorder characterised by signs and symptoms of intestinal obstruction in the absence of mechanical obstruction.¹ With no specific aetiology to tackle, management of CIPO focuses on symptom control and maintenance of an adequate nutritional status. Intractable chronic abdominal pain is a frequent and distressing feature in CIPO.

Here we report on the effect of bilateral thoracoscopic splanchnicectomy (TS) in two boys, one mentally retarded, with severe CIPO refractory to conventional therapy. Their debilitating chronic abdominal pain and markedly impaired quality of life (QoL) was iteratively alleviated by transient bilateral coeliac blocks (CB). After 26 and 9 successful CB, respectively, TS was performed. The greater and lesser splanchnic nerves were coagulated and divided bilaterally in both patients (fig 1).

The postoperative course was uneventful. Mild diarrhoea occurred on the first day after CB and was followed by a lasting improvement in bowel habits. Resolution of complaints and clinical signs, including a dramatic improvement in psychomotor and social behaviour of the mentally retarded boy, persisted over a follow up period of 30 and 25 months, respectively.

Management of CIPO remains challenging. Various therapeutic options have been proposed: dietary measures, total parenteral nutrition, prokinetic drugs, surgical procedures (for example, feeding gastrostomy or jejunostomy), gastrointestinal decompression procedures, gastric or intestinal pacing, bowel resections, and small bowel transplantation.^{2,3} Nevertheless, QoL often remains disappointingly poor. Chronic abdominal pain is a distressing feature in CIPO. Pain is especially difficult to assess in toddlers and in mentally retarded children. In teenagers, chronic pain is related to low self care and decreased participation in school and social activities.⁴

CB and TS both proved efficient for pain relief in cancer of the pancreas and in chronic pancreatitis.^{5–7} CB with local anaesthetic agents appears quite safe but offers only transient relief. Conversely, CB performed with destructive agents offers longer lasting relief but carries a risk of serious side effects, including paraplegia and circulatory arrest.^{8,9} We made the safe choice of anaesthetic CB for its reversibility as an unconventional therapeutic test. Transient diarrhoea, followed by improvement in bowel habits, was observed after CB and TS in both patients. This likely reflects relief of the gut from inhibitory sympathetic efferent signals, leading to an increase in motility and decrease in water absorption by excitatory cholinergic innervation.¹⁰ The role of the intrinsic enteric nervous system in the generation of excessive sympathetic reflex remains unclear. Improvement in gut motility after CB or TS suggests that, despite marked abnormalities (hyperganglionosis), the enteric nervous system was still able to organise adequate peristalsis after interruption of hyperactive sympathetic inhibitory reflexes. Furthermore, in both patients, immediate pain relief was observed after CB and TS, emphasising the prominent role of sympathetic afferent pathways in their painful symptoms. Altogether, these observations suggest that sympathetic reflexes played a central role in the physiopathology of painful CIPO in these two young patients.

The large number of procedures before TS (our patients underwent 26 and 9 CB, respectively) illustrates both the striking improvement provided by CB that strengthened treatment compliance and the reluctance of physicians to perform an irreversible procedure for an unconventional indication.

Based on our experience, we propose considering TS for children with refractory painful CIPO and poor QoL after a successful therapeutic trial with iterative CB. TS offers great

Figure 1 Thoracoscopic localisation and section of splanchnic nerves. Right pleural cavity, patient head on the right hand side. The greater (GS) and lesser (LS) splanchnic nerves were identified (A) and sectioned under visual control (B). D, diaphragm; V, intercostal vessels; L, lung.

promise for these patients and their families but still awaits dedicated investigations in larger series of patients to determine its place in the paediatric surgeon's armamentarium.

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Secondary amyloidosis in Crohn's disease: treatment with tumour necrosis factor inhibitor

We present the case of a 26 year old male patient diagnosed with ileocolonic Crohn's disease at age 15 years, with a history of several episodes of perianal inflammation that required surgical debridement. In 2000, he was hospitalised for tibio-malleolar oedema that had appeared progressively over the previous four months, associated with clinical signs of perianal inflammation and anal pain after defecating.

Physical exploration revealed a patient who was generally well, with a cushingoid appearance, pallor of skin and mucosae, oedema with fovea in both lower extremities up to the knee, minimum (not painful) fullness of the lower right quadrant and, on anal exploration, three uncomplicated "strings" with a suppurating point adjacent to one of them.

Laboratory tests showed $17\,720 \times 10^6$ leucocytes (11 600 neutrophils), haemoglobin 9.8 g/dl, platelets $577\,000 \times 10^6$ /l, cholesterol 294 mg/dl, triglycerides 197 mg/dl, total blood proteins 4.9 g/dl, albumin 2.4 g/dl, sedimentation velocity 122 mm,¹ β_2 -microglobulin 3.8 mg/l, urea 48 mg/dl, creatinine 1.7 mg/dl, fibrinogen 10.48 g/l, and negative antinuclear antibodies. He had an inactive urinary sediment, a creatinine clearance of 71 ml/dl, and proteinuria of 8 g/dl (selectivity index 0.22), which reached 20 g/24 h after a few days.

Abdominal Doppler ultrasound showed ileal disease, without local complications, and the small intestine follow through study showed two fixed and spiculated ileal loops. Renal ultrasound was normal.

The nephrotic range proteinuria in a patient with Crohn's disease made us suspect secondary amyloidosis. Colonoscopy was performed to evaluate intestinal inflammation as well as rectal biopsy to rule out amyloidosis. Rectal biopsy was positive for Red

Congo, which confirmed the suspected diagnosis (fig 1).

Treatment with Infliximab and azathioprine (100 mg/day) was indicated because of the severity of the perianal disease, which had not responded to conventional treatment. The patient received an anti-tumour necrosis factor α (TNF- α) infusion at weeks 0, 2, and 6, and every two months thereafter for four years. Colchicine is the empirical treatment for amyloidosis.

Perianal disease inflammation remitted completely and proteinuria began to decrease. Proteinuria values were 4 g/24 h after one year of treatment and 0.82 g/24 h after four years of treatment. The patient was treated with the three drugs (Infliximab, azathioprine, and colchicine) for the first four years and then with Infliximab for the last year (fifth year of treatment). Although asymptomatic, rectosigmoidoscopy to obtain new biopsies was performed after four and five years of treatment. The mucosa was macroscopically normal and biopsies were only slightly positive for Red Congo (there were a few irregularly distributed amyloid deposits) (fig 1) and positive on immunohistochemistry for substance P.²

To date, there has been no effective treatment for secondary amyloidosis^{3–5}; treatment consisted of treating the underlying inflammatory disease.^{2,6,7} This case is interesting because, when treated with Infliximab, there was an important clinical and analytical improvement, apparently followed by a decrease in the degree of Red Congo stain for amyloid deposits.

The clinical improvement in Crohn's perianal disease and the kidney abnormalities caused by amyloidosis (with an evident decrease in proteinuria values), as a response to the immunomodulating drugs, was remarkable, but even more so the fact that less amyloid deposits were seen in the second and third rectal biopsies.^{8,9}

Further study is required to confirm the relationship between treatment and clinical improvement¹⁰ and to determine if the combination of the three drugs is necessary (immunosuppressor (azathioprine) + immunomodulator (Infliximab), and an anti-inflammatory drug (colchicine)), or if

Figure 1 Red Congo stain before (A, year 2000) and after (B, year 2005) five years of treatment.