Abdominal emergencies

Vascular Ehlers-Danlos syndrome predisposes young adults to a high risk of digestive tract events which can be life-threatening. Bowel rupture is uncommon in early childhood, has been described in late childhood, and continues to be a risk into adulthood. Bowel rupture ultimately affects about 25–30% of individuals but rarely leads to death.

The main abdominal emergencies are (in order of severity):

- spontaneous arterial rupture in the peritoneal and/or retroperitoneal cavity,
- spontaneous digestive tract perforation, most often of the sigmoid colon, but also of the rest of the colon, the small intestine, and even the stomach,
- spontaneous rupture of an intra-abdominal organ (spleen, liver).

WHAT IS RECOMMENDED

- Perform an emergency angiography scan in the presence of an acute abdominal syndrome.
- In the event of colonic perforation, a wide colonic resection is the preferred method of treatment (Hartmann type for sigmoid colon).
- In cases of arterial rupture, percutaneous embolization should be preferred over open surgery.
- The use of stents should be limited to life-threatening emergencies, and embolization preferred whenever possible.
- In cases of spontaneous perforation of the digestive tract in a young adult or a child, a diagnosis of vascular Ehlers-Danlos syndrome should be automatically considered.
- A protocol of permissive hypotension is recommended without compromise to organ function.
- Caution with use of inotropes is recommended.
- Caution with use of indwelling catheters is advised.

Abdominal emergencies

WHAT YOU SHOULD NOT DO

- Attempt a minimal repair of a spontaneous perforation of the digestive tract.
- Perform a postoperative follow-up colonoscopy in the event of perforation of the digestive tract.
- Perform open surgery as first-line treatment in cases with arterial rupture.
- Any invasive or semi-invasive examination, particularly when the expected diagnostic rewards are minimal.