



Ehlers-Danlos Syndrome EDS

Emergency Physician's Reference CD-ROM



EDS Vascular (Type IV) Trauma Information



Physicians who have gained knowledge by treating the severe form of EDS



Updated information for the medical field



← Emergency

IF THIS IS A TRAUMA SITUATION, TIME IS OF THE ESSENCE!

Here is a condensed list of life-saving surgical and post-operative suggestions for patients with Ehlers-Danlos Syndrome - Vascular, Type IV. Although considered rare, clinical diagnosis of EDS Vascular is often difficult. In a trauma situation do not assume that your EDS patient has been typed correctly. EDS Vascular is a life-threatening connective tissue disorder that affects all tissue, arteries and internal organs making them extremely fragile.

Roughly 1/2 of all cases of Vascular EDS are new mutations with no family history. The other 1/2 are familial, inherited from an affected parent. Vascular EDS is autosomal dominant. Continue through the CD or booklet after this list, for more detailed information.

1. **CT scans or MRI's - immediately**
2. **No arteriographics, enemas, or endoscopies**
3. **Non-invasive techniques only - no stress/tension on skin, organs, or vessels - extreme care during physical exam or passing nasogastric tubes**
4. **Anesthesiologist please note: when intubating - fragile mucus membranes throughout - a lower peak volume pressure may be necessary**
5. **Vascular surgeon's assistance anticipated in every surgery - meticulous, gentle handling of internal organs, and vessels**

6. **Plastic surgeon's presence may be necessary**
7. **Aneurysm - a small soft tipped catheter with micro coil (memory) has been successful in some cases**
8. **Abdominal aneurysm - Double woven velour/Teflon grafts**
9. **Colonic rupture - consider permanent colostomy/ileostomy to reduce the risk of recurrent perforation**
10. **Padded clamps with red rubber catheter covers (Fogarty Hydrogrips)**
11. **Use Lange's lines for incisions - whenever possible (Teflon sutures)**
12. **Incision pressure - use 1/3 -to- 1/2 less pressure, with meticulous, gentle dissections - avoid tension/stress on suture lines.**
13. **Ligation of vessels - use surgical hemoclips and umbilical tapes - where anastomosis is required, buttressed sutures by Teflon or felt pledgets**
14. **If necessary the sacrifice of a non-essential organ or limb to save a life must be considered**

(Condensed Emergency Post Operative Care Suggestions - see page 3)

1. Monitor for: peritonitis, pneumoperitoneum, and/or other infections
2. Monitor for: ruptures, cysts, and abscesses
3. Monitor for: wound dehiscence, ileus, gastrointestinal bleeding
4. Monitor for: arteriovenous and/or intestinal fistula
5. Monitor for: aneurysms, embolus, hematoma
6. Monitor for: eventration of diaphragm, pleural effusion, pneumothorax
7. Monitor liver for: bleeding, changes in pressure and/or function
8. Wound packs and abdominal binders (reduce risk of incisional hernia)
9. Monitor for: increased or erratic blood pressure
10. IV placement: may be problematic due to fragile veins (If necessary, permanent access port catheter has been used)
11. Less IV pressure: slower rate when administering fluids
12. Immediate evaluation - of any change in vitals or additional complaints
13. The most non-invasive post-operative care available is recommended
14. Be vigilant - as status can change abruptly with this patient

POSSIBLE HOLLOW VISCUS RUPTURE:

“Defective wickerwork of collagen”, the result of an inability of the abnormal collagen fibrils to organize into bundles that are essential for the formation of a strong network”. (Jansen 1955) “This results in defective type III collagen, a protein that’s expressed in many tissues but is primarily a component of extensible connective tissues such as skin, gastrointestinal tract, bladder, uterus, the highly cellular structures such as liver, lungs, and vascular system.” (Dagleish 1998)

INDICATIONS AND SUGGESTIONS:

“Acute abdominal, chest, inguinal, and flank pain (diffuse or localized) is a common presentation of arterial or intestinal rupture and should be investigated urgently. Non-invasive diagnostic procedures are recommended.” (Beighton et al. 1998)

“Enemas - should not be used because of colonic distention that may result in perforation.” “Endoscopy -” Although it has been performed without complications in a limited number of patients, it should be avoided.” (Solomon et al 1996)

“Doppler ultrasound, duplex or computed tomography(CT) scan, transesophageal echocardiogram(TEE), magnetic resonance or intravenous digital subtraction angiography (DSA) should be used.” (Karkos et al. 2000) & (Sherry et al.1992)

“Utilize invasive technique only when the information it provides is essential for pre-or intraoperative decision making.” (Whitehill et al. 1995, Brearley et al. 1993, Barabas et al. 1990, Cikrit et al. 1987) *Read important quotes concerning “Arteriography”, listed under POSSIBLE ARTERIAL RUPTURE & SURGICAL TECHNIQUES

“Early diagnosis of peritonitis is critical so the appropriate therapy can be instituted expeditiously, including correction of fluid and electrolyte abnormalities, institution of antibiotic therapy, and surgical repair of the underlying lesion.” (Kinnane et.al 1995)

“Sepsis/septic shock: Early recognition is needed to prevent acceleration, as the microcirculation undergoes massive alteration.” (Hinshaw 1996)

“Spontaneous colon perforations are most commonly reported in the sigmoid colon, but can also occur anywhere in the colon and rectum. A history of constipation may precede perforation.” (Solomon et al. 1996)

“Prompt surgical intervention was normally crucial in the treatment of bowel rupture, and colostomy was the preferred treatment.” (Pepin et al. 2000)

“After a colostomy is placed, stool softeners remain an important therapy as well as careful surveillance for evidence of colostomy stenosis.” (Solomon et al. 1996)

Latest clinical/genetic research on the Vascular Type - the study indicates: “Bowel continuity was restored with little difficulty in most cases. Treatment of bowel perforation with end-to-end reanastomosis after partial colectomy was associated with a higher risk of both immediate failure and later complication than was treatment with colostomy. The sigmoid colon is a frequent site of rupture, removal of the distal colon may decrease the risk of recurrence.” (Pepin et al. 2000)

“Restoration of the colostomy cannot be considered curative, as other perforations can recur, especially if there is evidence of colostomy stenosis, complications can arise from the breakdown of the anastomosis, and death as a result of this breakdown has been reported.” (Stillman et al. 1991, Solomon et al. 1996)

“The safest procedure is considered to be a total colectomy and/or a permanent ileostomy.” (Stillman et al. 1991)

“Small bowel ruptures have also occurred, these have been managed by resection and performing end-to-end anastomoses, without complication.” (Solomon et al. 1996)

“Dilation of the entire small bowel from the ligament of Trietz to the terminal ileum has been observed without evidence of malabsorption or bacterial overgrowth.” (Harris 1974)

“Intramural hematomas may then cause focal areas of necrosis in the bowel wall, leading to perforation.” (Solomon et al. 1996)

“The loss of strength of connective tissue may interfere with the ability to wall off infectious processes and therefore increase the risk of abscess formation leading to increased morbidity and mortality when perforations occur.” (Solomon et al. 1996)

“Colonic fistulas, colostomy breakdown, hematoma formation, vascular accidents, prolonged bleeding, and multiple intraperitoneal adhesions, abscesses, enterocutaneous fistulas, may occur. Careful handling of abdominal contents intraoperatively, as surgical procedures in EDS patients may be problematic.” (Berney et al. 1994, Silva et.al 1986, Solomon et.al 1996)

“Pregnancy/Obstetrical complication: include ruptures of the uterus, premature rupture of membranes, rupture of blood vessels, aorta, vena cava, and gravid uterus, sigmoid colon tearing of perineum, vagina, urethra and bladder and sigmoid colon.” (Peaceman & Cruikshank 1987) (DePaepe et al. 1989) “Bladder ruptures due to vesical diverticula.” (Bachiller et al. 2000)

POSSIBLE ARTERIAL RUPTURE/ANEURYSM/HEMATOMA:

“Although operative mortality remains at a high level due to the tendency of vessels to tear with even minimal manipulation, mortality from hemorrhage without surgical intervention is even greater.” (Mattar et.al 1994)

INDICATIONS AND SUGGESTIONS:

“Presentation of abdominal, chest, or pelvic pain of a sudden onset - is suggestive of a vascular or hollow viscus catastrophe. To help prevent traumatic perforations, extreme care should be taken when performing physical examination, surgery, during placement of an IV, and passing nasogastric tubes.” (Solomon et al. 1996)

“Vascular complications often present as life-threatening emergencies and there may be no time for diagnostic studies to be undertaken.” Patients are frequently unaware of the disorder until sudden rupture of an artery or the bowel occurs. Rupture of the aorta is a catastrophic event that usually occurs spontaneously in Vascular EDS and is nearly 100% fatal.”(Karkos et al., 2000)

“Doppler ultrasound, duplex or computed tomography(CT) scan, transesophageal echocardiogram(TEE), magnetic resonance or intravenous digital subtraction angiography (DSA) should be used.” (Karkos.et al. 2000) & (Sherry et al. 1992)

“Arteriography” - “Reported complications: arterial laceration, false aneurysm formation, hemorrhage, arteriovenous fistulas, and death.” As a result, some authors believe that angiography is contraindicated.” (Sherry et al. 1992) “However, multiple successful uncomplicated attempts at angiography have been reported.”(Sherry et al. 1992, Mattar et al. 1994, Mirza et al. 1979) *Read more important quotes under: **SURGICAL TECHNIQUES**

“If arteriography is considered absolutely necessary, very fine catheters and a careful, and atraumatic technique are mandatory.” (Karkos et al. 2000)

“Vascular complications that have been reported: aneurysms, rupture, dissection, varicosities, bloodclots, and arteriovenous fistula formation of the; Aorta, abdominal, celiac, carotid, cerebral, iliac, subclavian, inferior/superior vena cava, epigastric, infra-renal, inferior/superior mesenteric, hepatic artery, portal vein, splenic, and pulmonary.” (Solomon et al. 1996)

“Cardiac problems may occur - symptoms of myocardial infarction warrant investigation of possible coronary artery dissection or tear - consider aortic root or coronary artery problems.” (Ades et.al 1995) (Evans & Fraser 1996)

“Preferred operative treatment is ligation of vessels, followed by bypass grafting only when necessary. Graft (double woven velour graft and Teflon) - for abdominal aneurysm, has been used successfully for repair.” (Mattar et.al 1994)

“Spontaneous arterial rupture has a peak incidence in the third or fourth decade of life but may occur earlier. Midsized arteries are most commonly involved. Arterial rupture is the most common cause of sudden death.” (Pepin et al., 2000)

“DDAVP (Prophylactic desmopressin) has helped to control bleeding in EDS.” (Stine KC, Becton DL., 1997)

“Pregnancy- Greatest risk for arterial rupture during perinatal period - labor and postpartum. Sudden pain or evidence of blood loss should be investigated promptly.” (Peaceman & Cruikshank 1987)

“Aside from vascular fragility, another unfortunate factor associated with hemorrhage is the lack of adjacent connective tissue structure to tamponade.” (Rybka & O’Hara 1967)

“In cases where surgery is deemed too risky, dissections have been managed successfully with antihypertensive therapy - This therapy is also mentioned for treatment of some aortic dissections that are inoperable.” (Gertsh et.al 1986, Solomon et.al 1996)

“Blood pressure should be monitored in these patients and any physical exercise in which a sudden increase of blood pressure is likely, must be avoided.” (Burrows, **The Management of EDS**)

“Intracranial aneurysms associated with the Vascular type, should be considered in the differential diagnosis of cerebrovascular disorders and stroke in early childhood.” (Kato et al 2001)

SURGICAL TECHNIQUES:

“The key to favorable outcomes lies in identification of the syndrome pre-operatively, to avoid the catastrophic vascular complications, surgical intervention only in life- or limb-threatening situations, and appropriate modification of surgical technique.” (Mattar et.al 1994)

“The Vascular Type of EDS is “dramatic, deadly and deceptive.” (Sparkman, 1984)

INDICATIONS AND SUGGESTIONS:

“Alert anesthesiologist to the potential mechanical complications associated with administration of anesthesia - hyperextension of the jaw during intubation should be done very gently to avoid trauma to the skin and deeper tissues.” (Wesley et.al 1980)

If surgery is indicated, a Vascular Surgeon should be in attendance. A plastic surgeon’s presence may be necessary.

“Techniques have to be adjusted - every pull on the patient’s tissues, and every cut with the scalpel, will require only about half or a third of the usual force used for a normal person. Meticulous, and gentle dissections - No stress or tension should be put on the skin, internal organs, blood vessels or the arteries.” (Barabas, **The Management of EDS**)

“Angiography” - there have been reports of complications: (See **POSSIBLE ARTERIAL RUPTURE**) “As a result, some authors believe that angiography is contraindicated.” (Sherry et al. 1992) “However, multiple successful uncomplicated attempts have been reported.” (Sherry et al. 1992, Mattar et al. 1994, Mirza et al. 1979)

“In an effort to embolize the fistula, four 5mm Gianturco coils were placed into the proximal proper hepatic artery. There were no complications during angiography.” (Sherry et al. 1992, Noshier et al. 1986)

“Endovascular therapy of arteriovenous fistula, Carotid Cavernous Fistula (CCF) was treated by transvenous occlusion with regular and fiber-coated Guglielmi (electrolytically) detachable coils.” (Janson et.al 1999)

“Unless a life-saving procedure is needed, we believe that laparotomy should be avoided at all costs in these patients.” (Berney et al. 1994)

“Skin - bruising, scarring, wound dehiscence is a problem. On the skin there are naturally occurring linear marks which run in a specific direction, these are known anatomically as “Lange’s lines”. More problems seem to occur when the surgical incision has been made across this grain. Therefore, if the incision is made in the same direction as the naturally occurring lines, then there is a good possibility there will be less scarring, less pain, and better healing. Use surgical instruments that are lighter than usual, such as retractors and

blood vessel clamps. Padded clamps with red rubber catheter covers or Fogarty Hydrogrips should be used.” (Bunt & Malone 1993, Mattar et.al 1994, Barbabas **The Management of EDS**)

“Arteries may tear when conventional clamps are applied using normal amount of force. Soft peripheral arterial, rather than heavy aortic clamps, should be used to occlude the aorta, but aortic occlusion should be used with extreme care.” (Whitehill 1995, Barabas 1990, Karkos et al 2000)

“DDAVP (Prophylactic desmopressin) has helped to control bleeding in EDS.” (Stine KC, Becton DL., 1997)

“Preferred operative treatment is ligation of the arteries with sutures or bypass grafts only when necessary . In cases of rupture of middle-sized arteries, the patient’s life may be saved by sacrificing a nonessential organ or a limb.” (Karkos et.al 2000) Graft (double woven velour graft and/or Teflon) - for abdominal aneurysm, has been used successfully for repair.” (Mattar et.al 1994)

“Ligation of vessels should “not” be performed with sutures - because they tear through the fragile vascular walls. Using umbilical tapes and surgical hemoclips just proximal to the site of bleeding.” Another unfortunate factor may be the lack of adjacent connective tissue structure to tamponade.” (Mattar et.al 1994)

“If vessel anastomosis is required for reconstruction, interrupted, horizontal mattress sutures buttressed by Teflon or felt pledgets are suggested.” (Mattar et.al 1994)

“Surgical management of the EDS patient is very delicate and hazardous.” (Karkos et al 2000) “Surgeons who have had the unfortunate privilege of operating on such patients describe the extreme friability of the tissues and vessels encountered. The tissue are

described as resembling “wet blotting paper”, “cold porridge”, or “wet cotton”, while the arteries and veins tear easily because of the flimsiness of their walls, literally crumbling in the surgeon’s hands. “Normal” handling of bleeding arteries and tissue usually induces new ruptures, tears or small hematomas.” (Karkos et al. 2000) & (Schievink et al., 1990)

“Tremendous intraperitoneal adhesions and densely adherent colon and abdominal tissues may be encountered, probably caused by small perforations which seal off against the surrounding tissue which, when associated with intestinal fragility, cause surgery to be laborious and frustrating.” (Berney et al. 1994, Sykes 1984)

“Stitches should be made in all layers, ligatures must not be pulled tight but gently approximated. Keep stitches in place for at least twice as long as normal. Teflon sutures, reinforced with surgical tapes, such as wide steri-strips longitudinally (without tensing the skin), have been shown to be more effective on very fragile tissue.” (Barbabas & Attwood, **The Management of EDS**) & (Mattar et.al 1994)

POST OPERATIVE CARE:

“The approach to the EDS patient needs to be individualized; differentials should be broad, and consideration must be given to the potential for complications if invasive procedures are performed.” (Solomon et al. 1996)

INDICATIONS AND SUGGESTIONS:

“With Vascular EDS give immediate attention, be aware of the severity of the situation - stay alert for other potential problems that could occur. Unacceptable delays could mean the difference between life and death.” (Karkos et.al., 2000)

“Sepsis/septic shock: Early recognition is needed to prevent acceleration. As the microcirculation undergoes massive alteration, which may lead to hemorrhagic necrosis terminating in shock and death.” (Hinshaw 1996)

“Blood pressure should be monitored in these patients and any physical exercise in which a sudden increase of blood pressure is likely, must be avoided.” (Burrows, *The Management of EDS*)

“Check coagulation, platelets for bleeding disorders” - “Most laboratory studies reveal bleeding and clotting times to be normal, even though most require blood transfusions and intravenous alimentation. Recent studies have shown that the increased bleeding tendency probably is due to a defect in the collagen structure of the vascular and perivascular tissues. There is also a reduction in the ability of abnormal collagen in patients with EDS to attract platelets, which may contribute to the bleeding tendency.” (Karaca et.al 1972) (Wesley et.al 1980)

“Nonoperative management with bed rest, monitoring conservatively, and external compression, elevation of extremity, if feasible, may be the optimal treatment for bleeding of peripheral vessels.” (Whitehill 1995, Barabas 1990, Cikrit et.al 1987, Karkos et.al 2000)

“Pressure therapy is used to diminish dissection of the arterial hemorrhage into the surrounding tissues. Observe area for tactile rippling up and down the involved extremity, if unimpeded, can avulse the small vessels feeding the tissues superficial to the level of the hemorrhage, leading to necrosis and slough of these overlying tissues.” (Wesley et.al 1980)

“If extremity pressure therapy fails, ligation of major bleeding vessel just proximal to the site of bleeding is necessary using great care not to accidentally divide the vessel and use

very gentle techniques and heavy suture material, umbilical tape is suitable for this purpose.” (Wesley et.al 1980)

“The use of stay sutures at a distance from the wound as well as the use of measures to decrease intraabdominal pressure by preventing cough, ileus, and bladder outlet obstruction have been advised for prophylaxis against dehiscence.” (Stillman et. al 1991) (Solomon et.al 1996)

“Prolonged oozing from all traumatized surfaces, have been reported.” (Soucy 1990)

“Post-operative period, EDS patients are prone to certain complications: due to the low tissue strength and poor wound healing, wound dehiscence and incisional hernias may occur. In these situations, the use of wound packs and abdominal binders has been recommended.” (Sykes, 1984, Solomon et al, 1996)

“High rate of wound infection has also been reported. The propensity for poor healing and dehiscence.” (Beighton et.al 1969, Solomon et.al 1996)

The usual care should be taken to monitor for postsurgical complications: “Individuals with the Vascular type have been reported with: “infections, sepsis, wound dehiscence, ileus, severe gastrointestinal bleeding, arteriovenous fistula, aneurysms, hematoma, pneumothorax, embolus, pleural effusion, hemoptysis, tremendous intraperitoneal adhesions, scar tissue, rectal prolapse, pneumoperitoneum, paraesophageal hiatus hernia, eventration of the diaphragm, hernias, cysts, abscesses.” (Wesley et.al 1980)(Solomon et.al 1996)

“Microangiopathy of the skin capillaries with microbleedings, presence of microaneurysms and increased transcapillary diffusion. Microvascular involvement appears to be an additional manifestation of the syndrome.” (Superti-Furga et al. 1992)

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Please Note:

Although Vascular Ehlers Danlos Syndrome - Type IV is rare, the possibility of complications with all forms of EDS should be considered in a surgery or a trauma situation. Do not assume that your patient has been typed correctly, as clinical diagnosis is often difficult. Take necessary precautions during procedures with any type of EDS.

Minimum System Requirements

Autorun	Autorun
Windows 95/98, Win NT	Mac OS 7.5.3 or later
64 MB RAM, 16 bit color	64 MB, 16 bit color
4x CD-ROM	4x CD-ROM
Pentium II recommended	Power PC

INSTRUCTIONS: This CD is autorun with nothing to load on the user's computer. Simply insert the CD in your CD drive and navigate to a chosen topic. If you are having trouble playing the CD, reboot your system. Let the Introduction play through completely. Click into the Trauma section and then let the initial video in that section play through completely before selecting another section.

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