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### Case Report

# Successful management of superior mesenteric artery aneurysm in a 16y old boy: Case report

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### Abstract...

Ehler`S -Danlos Syndrome (EDS) is a challenging inherited collagen disorder, characterized by joint hypermobility, increased skin elasticity and tissue fragility. It is a familial disease. 50% of diagnosed patients have sporadic new mutations without any antecedent familial history. EDS is classified into 13 genotype-based subtypes. Although subtype I and II are the most common, subtype IV EDS which has a unique COL3A1 gene mutation coding type III procollagen and known as vascular subtype because of the many vascular complications including: spontaneous or induced arterial dissection, aneurysm, pseudo-aneurysm and spontaneous arterial rupture. This subtype has the lowest survival and highest mortality rate. Patients with nearly any subtype are also prone to vascular manifestations. Management of these complications include: observation in asymptomatic and non life threatening ones, open surgical ligation or endovascular coiling of the affected artery and finally arterialre-construction (grafts\stent-grafts) when organ vitality is threat end. We present a case report of successful symptomatic Superior Mesenteric Artery Saccular Aneurysm (SMAA) repair in a-16- year old boy, previously diagnosed with EDS, treated by a stent graft and underwent a normal 16month's follow-up. A literature review of these cases regarding their management especially by endovascular procedures would be considered.

**Keywords:** Ehler`S -Danlos Syndrome (EDS); Collagen disorder; Superior mesenteric artery aneurysm; Stent graft.

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## Introduction

Ehler's -Danlos Syndrome (EDS) is one of the inherited connective tissue disorders, with an estimated prevalence of 1/50,000-90,000. Based on signs and symptoms it is classified into seven major clinical groups having common characteristics of joint hyper mobility, skin hyper-extensibility, and tissue fragility affecting the skin, ligaments, joints, blood vessels, and internal organs. The natural history of each type differs between these groups what makes it important to specify each patient based on clinical, biochemical and genetic examinations. Although groups I and II are the most common, group IV known as vascular EDS accounts for less than 5%, has the highest catastrophic vascular complications (arterial rupture) and the lowest life expectancy. Though rare, in childhood its onset increases with time, and at least 80% of patients may have had a complication during their life. Our knowledge of EDS is mainly based on reports of small samples and few case reports showing that vascular manifestations were most commonly noted at the thoracic and abdominal aorta, and renal arteries were the most affected visceral arteries. Many patients are asymptomatic and incidentally detected. Symptomatic vascular complication management is vital though challenging in view of generalized tissue fragility. Artery ligation, traditionally, was the first therapeutic approach but led to catastrophic organ's ischemia, life-threatening complications and death. Early reconstruction attempts were successful, yet ligation remained useful especially in recurrent aneurysms, anastomotic site complications, and life-threatening rupture. Endovascular procedures made arterial reconstruction or coiling possible with lower complication rates. The continuous endovascular devices development and the accumulative experience led to successful endograft reconstruction without devastating complications.

We present a successful endovascular repair of symptomatic superior mesenteric artery aneurysm with a literature review of EDS's visceral artery complication management.

## Case presentation

A 16 years old boy presented with 10 months history of episodic abdominal cramps and pain, not related to meals, and previously managed only with analgesics. The last episode started 15 days before admission was non-responsive to analgesics at all. The young boy was already known to have EDS based on his clinical aspect: tendency to easy bruising, hyper mobility of joints and fingers, and kyphosis (Figure 1). He was the first of four family members to suffer EDS, his 12 years old sister and two cousins (8 years -10 years old) were later diagnosed with EDS at birth depending on familial history and physical examination alone. The abdominal palpation showed peri-umbilical pain without any abdominal parietal rigidity. Hemoglobin level was 13.7 g/dl and leukocyte count was 9.64 k/ml. A urine sample was normal. Abdominal ultrasound evidenced a 30mm antero-posterior diameter aneurysm of the Superior Mesenteric Artery (SMAA). Contrast abdominal Multi-Slice Computed Tomography (MS-CT) confirmed 25 x 30 mm superior mesenteric artery aneurysm, starting 15 mm distal to its origin from the abdominal aorta (Figure 2). Under general anesthesia, an eight French sheath introduced into the right common femoral artery percutaneously. A 0.035-inch guide wire and a 6 French simon I catheter engaged selectively the SMA. Subsequently, a 0.035

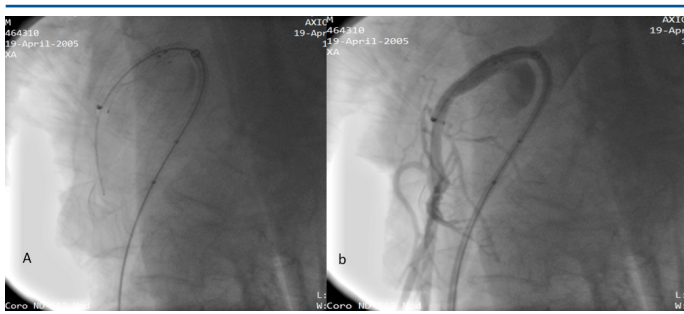
inch Amplatz extra-stiff wire was gently placed distally in the SMA and a 7 x 30 mm stent-graft (FLUENCY plus Vascular Stent Graft) across Lifetech Fustar steerable introducer was deployed to exclude the aneurysm. The control angiography showed leakage at the proximal neck totally excluded by a 7 x 19 mm Balloon-Expandable Stents (CID-isthmus) (Figure 3). A color duplex and contrast-enhanced CT 10 days after endovascular treatment confirmed a patent SMA and total thrombosis of the aneurysmal sac without any leakage (Figure 4).



**Figure 1:** Patient characteristics, (A): general appearance note the spontaneous diastasis recti (blue arrow). (B,C): Hypermobility of small joints, acrogeria and thin translucent skin. (D): easily bruising.



**Figure 2:** Computed tomographic angiography scan of the abdomen showing a superior mesenteric artery aneurysm. (A) Axial section. (B) Coronal section. (C) Sagittal section.



**Figure 3:** Intra-procedural angiogram. (A): stent-graft is deployed. (B): An endo-leak type I, and deploying the stent to close it is (arrow).



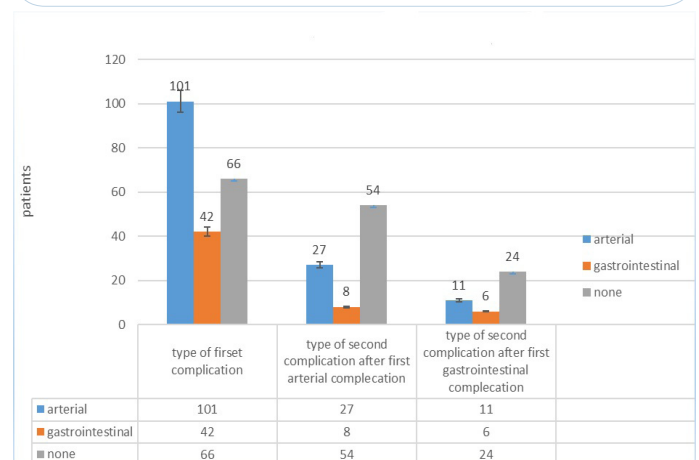
**Figure 4:** Computed tomography scan 10 days after endovascular treatment confirming technical success. The aneurysm completely excluded, and patency of the superior mesenteric artery is preserved.

## Discussion

EDS's Vascular complications management are not standardized due to the rarity of the disease. There is an accepted consensus for conservative management when dealing with asymptomatic EDS, with follow-up (every 2-3 years). Patients younger than 20 years old optimal follow up is not yet established [4]. Individualizing treatment for symptomatic patients by surgical or endovascular approaches is vitally important. Many experts dealing with such sophisticated rare disease prefer the "hit-and-run" concept, by ligating or coiling the affected artery if this does not compromise the organ or limb vitality. This is because of the high risk of bleeding and anastomotic site complications associated with surgery (37% and 40% respectively according to Oderich) [3]. Endovascular procedures gained much confidence -nowadays- as an alternative management for these patients despite the initial fear of puncture site complications, iatrogenic

rupture during maneuvers due to arterial fragility, and the possibility of compromising collateral circulation. The continuous development of new low-profile devices made it possible to operate with minor complications compared to open surgery. The largest cohort of 419 subjects by Pepin et al. [5] identified 272 arterial complications; noticing that arterial complication was the most common first and second complication (46% and 30% respectively) (Table 1), whilst, 92% of late deaths were due to vascular complications. Reviewing 31 patients with vascular EDS at Mayo Clinic (1971-2001) Oderich [3], showed that the most detected vascular complications were at thoracic and abdominal aorta (10 and 7 patients respectively), while the visceral complications affected renal arteries most commonly, followed by Superior Mesenteric Artery (SMA) (nine, six patients respectively); then hepatic and celiac arteries equally (four patients for each), one case involved the inferior mesenteric artery. Only eight out of 183 patients needed arterial different procedures at Johns Hopkins Hospital. Viscerally a hepatic pseudoaneurysm and two splenic aneurysms were treated with coil embolization, gel foam was needed in one of the splenic; another visceral manifestation was reported as renal artery stenosis treated by balloon-expandable stent was complicated by dissection and needed another stent [1]. Renal artery aneurysm repair in two patients affected by EDS (one true and one pseudoaneurysm) by stent-graft, showed thrombosis after 43 months follow-up in one patient, and full patency in the other [6]. Four other case reports were conducted for the management of EDS's visceral complications. Three SMA spontaneous aneurysms were managed by stentgrafts. Leeuw et al., [7] presented a 9 years old boy case report who was treated by a stent graft for SMAA which then complicated by rupture and underwent emergent laparotomy ligating the SMA which led to intestinal ischemia and death of the child after 9 days. Schweigert et al. [8] Yasuda et al. [9] treated a 13 years old female and 79 years old male respectively by stent grafts for SMAA without any complication. The fourth case report was of an inferior mesenteric artery IMA aneurysm treated successfully by coiling and foam embolization to achieve total occlusion without any complications Rahman et al., [10].

**Table 1:** Complications in 220 patients with vascular EDS, and the relation between the second and first complication.





**Table 2:** Summarizes EDS's visceral arteries complications and ways of management. Literature review of EDS's visceral arteries complications, management and outcomes. N=14

| Author\year number of patients | Artery\number of affected arteries | Type of complication                              | Management                            | Follow-up   |
|--------------------------------|------------------------------------|---|---------------------------------------|---|
| Oderich 2005 n=4*              | Hepatic n=1                        | Ruptured aneurysm                                 | Interposition graft                   | Died at 62 y-aortic rupture                                       |
|                                | Splenic n=2                        | Ruptured aneurysm                                 | Splenectomy                           | Died at 62 y-aortic rupture                                       |
|                                |                                    | Ruptured pseudoaneurysm                           | Splenectomy                           | Died at 44y- arterial rupture                                     |
|                                | Inferior epigastric n=1            | Spontaneous rupture                               | Coil embolization                     | -----   |
| Lum YW 2011 n=4*               | Ileocolic n=1                      | Spontaneous rupture                               | Primary ligation                      | Died at 84y –arterial rupture                                     |
|                                | Hepatic n=1                        | pseudoaneurysm                                    | Coil embolization                     | -----   |
|                                |                                    | Aneurysm  | Coil embolization                     | Liver hematom +hemoperitoneum after 2 y                           |
|                                | Splenic n=2                        | Aneurysm → progressed into 4 aneurysms within 4 y | Coil+gelfom embolization              | No complications  |
| Künzle 2013 n=2                | Renal n=2                          | Stenosis then dissected after stent deployment    | Peripheral stent (balloon expandable) | The only complicated patient which was treated with another stent |
|                                |                                    | Aneurysm  | Stent graft                           | Thrombosed after 43 m   |
| Schweigert 2011 Case report    | SMA n=1                            | pseudoaneurysm                                    | Stent graft                           | Patent  |
| De leeuw 2011 Case report      | SMA n=1                            | Aneurysm  | Stent graft                           | Patent, no endoleak   |
| Yasuda 2013 Case report        | SMA n=1                            | aneurysm  | Stent graft                           | Rupture of the aneurysm → emergency laparotomy and ligation       |
| Rahman 2019 Case report        | IMA n=1                            | Spontaneous rupture                               | Stent graft                           | Patent for 2 y  |
|                                |                                    | aneurysm  | Coil+foam embolization                | Occluded for 1y with no complications                             |

### Conclusion

Endovascular stent-graft placement was successful in our case and it may represent a low-risk option for managing vascular complications in selected patients with type IV EDS.

### Compliance with ethical standers

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**Availability of data:** The datasets collected and/or analyzed during the current study are available from the corresponding author on request. The corresponding author had full access to all the data in the study, and takes the responsibility for the integrity of the data and the accuracy of the data analysis.

**Ethics approval and consent to participate:** All the performed procedures were in accordance with the ethical standards of our institutional research committee, and with the 1964 Helsinki declaration and its later amendments. A written informed consent has been obtained from our patient.

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