

1 **Factors influencing the recurrence of arterial involvement after**
2 **surgical repair in Behçet's disease**

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20 This study was presented in the plenary session at the 2019 Vascular Annual Meeting of the
21 French Society for Vascular and Endovascular Surgery, Lille, June 28-30, 2019.

22 This study has been selected for fast-track presentation at the 2019 Vascular Annual Meeting
23 of the European Society for Vascular and Endovascular Surgery, Hamburg, September 24-27,
24 2019.

26 **ARTICLE HIGHLIGHTS**

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28 **Type of Research:** single center retrospective study

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30 **Key Findings:** Twenty-three patients with BD, had aortic and peripheral arterial repair between
31 May 1996 and September 2015. Twenty-four recurrences were noted and forty-seven surgical
32 procedures were performed. Mean follow-up was 8.4 ± 7.5 years. Initial arterial lesions were
33 aneurysms and thrombosis in 85% and 15% of cases, respectively. Recurrence rate was 51%.
34 Preoperative medical treatments, including colchicine, steroids or immunosuppressants,
35 significantly decreased recurrence rate: 28% (7/25) *versus* 75% (15/20) in untreated patients (P
36 = 0.002). When anastomoses were protected using a prosthetic sleeving technique, the
37 recurrence rate was threefold lower (P = 0.08).

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40 **Take home Message:** Vascular Behçet's disease is responsible for a high rate of recurrence
41 after surgical repair. Perioperative immunosuppressants are essential to reduce this risk. The
42 additional surgical technique consisting in sleeving anastomosis may help to reduce
43 pseudoaneurysm recurrences occurring at the same site in 92% of cases.

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46 **Table of Contents Summary**

47

48 Recurrence after surgical repair occurred in 51% of cases in this retrospective study of 23
49 patients with non-pulmonary arterial involvement of BD. Immunosuppressive treatment but
50 also mechanical sleeving of the anastomosis decrease recurrence rates.

52 **Abstract**

53 **Introduction:** Arterial involvement in Behçet's disease (BD) is rare and its surgical
54 management is a major concern because of its high recurrence rate. This study evaluated the
55 influence of the surgical technique, device, and immunosuppressive treatment used on the
56 postoperative recurrence in patients with non-pulmonary arterial BD.

57 **Methods:** Single center retrospective study conducted in 23 patients meeting the International
58 Study Group of BD criteria, who underwent surgery for arterial involvement between May 1996
59 and September 2015. Recurrence was defined as the occurrence of arterial aneurysm or
60 thrombosis during follow-up. Perioperative medical treatment and surgical technique used were
61 reported.

62 **Results:** Forty-seven surgical procedures were performed in 23 patients. Mean follow-up was
63 8.4 ± 7.5 years. Initial arterial lesions were aneurysms and thrombosis in 85% and 15% of cases,
64 respectively. Arterial lesions were aortic and peripheral in 48% and 52% of cases. Recurrence
65 rate was 51%. Recurrences occurred within less than one year in 24% of cases and at the same
66 anatomical site in 92% of cases. Among the 24 recurrences, 17 were false aneurysms, 6 were
67 thrombosis and one was a true aneurysm in a different arterial site. To treat the arterial lesion,
68 direct anastomosis was performed in 6 cases, bypass using the saphenous vein, graft or allograft
69 was performed in 6, 27 and 5 cases, respectively, and stentgraft was used in 3 cases. Vascular
70 lesions involved the aorta in 19 cases and a peripheral artery in 28 cases.

71 Preoperative medical treatments, including colchicine, steroids or immunosuppressants,
72 significantly decreased recurrence rate: 28% (7/25) *versus* 75% (15/20) in untreated patients (P
73 = 0.002). The recurrence rate was 42.5% (17/40) in patients treated postoperatively *versus* 80%
74 (4/5) in untreated patients. The nature of the device used (vein, prosthetic graft, allograft, stent
75 graft or direct anastomosis) did not change the risk of recurrence. When anastomoses were

76 protected using the prosthetic sleeving technique, the recurrence rate was three times lower (P
77 = 0.08).

78 **Conclusion:** Relapse is a main concern after surgical repair of arterial BD. This study suggests
79 the need for targeted perioperative medical management to reduce the risk of arterial recurrence
80 in BD patients. To this end, a multidisciplinary approach is mandatory.

81 The use of sleeve anastomosis is associated with a numerically lower risk of recurrence.

82 However, further studies are needed to confirm this efficacy.

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85 **Key words:** Vascular Behçet's disease, aneurysm, recurrence, sleeve anastomosis.

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89 **Conflict of interest:** None

90

91 **Introduction**

92 Behçet's disease (BD) is a chronic, relapsing, multisystemic disorder characterized by
93 mucocutaneous, ocular, vascular and central nervous system manifestations^{1, 2}. The
94 pathogenesis of the disease remains unknown, although a genetic predisposition, environmental
95 factors, and immunological abnormalities have been reported^{3, 4}. Vasculitis is thought to
96 underlie the clinical manifestations of BD and is distinguishable because both arteries and veins
97 of all sizes are involved. Arterial complications are less common than venous lesions in BD,
98 and occur in 1-14% of patients⁵⁻⁸. Vascular BD (VBD) patients are at risk of multiple vessel-
99 related complications resulting in an increased risk of mortality. Indeed, the 20-year survival
100 rate decreases from 89 to 73% in case of arterial disease⁹. In a large cohort of 817 BD patients,
101 Saadoun et al.⁸ have shown an overall mortality rate of 5% after a median follow-up of 7.7
102 years. In this series, the incidence of arterial involvement was 3 times higher in patients who
103 died. Unlike venous manifestations¹⁰⁻¹⁵, arterial involvement in BD is rare and a few
104 publications have reported its specific treatment. Vascular surgery is challenging in BD patients
105 and the mortality rate is high¹⁶, mainly due to anastomotic aneurysm relapses and graft
106 thrombosis.

107 There is currently no consensual technique for the surgical management of these patients. The
108 vascular substitutes that can be used include prosthetic materials, venous autografts, arterial
109 allografts or stent grafts⁹. The main concern in these patients, apart from bypass thrombosis, is
110 the formation of anastomotic pseudoaneurysms in the short term, requiring iterative
111 interventions that may be life-threatening in case of rupture.

112 The aim of this study was to retrospectively identify the factors influencing the postoperative
113 recurrence of arterial lesions in BD patients with non-pulmonary arterial involvement.

114

115 **Patients and methods**

116 Population

117 It was a single center retrospective study conducted in 23 BD patients meeting the international
118 criteria for BD¹⁷ who consulted the Departments of vascular surgery and internal medicine of
119 the Pitié-Salpêtrière Hospital, Paris, France between May 1996 and September 2015.

120 Associated clinical signs were genital and oral ulcerations, uveitis, folliculitis, arthralgia, and
121 fever. Other aneurysmal locations, a history of deep vein thrombosis and cardiovascular risk
122 factors were identified. Pre- and post-operative medical treatments were recorded. Patients with
123 BD requiring arterial repair in our department were systematically included in the study whether
124 it was a recurrence or a primary surgery. In case of previous arterial repair in another center,
125 the first surgical repair in our department was considered a recurrence. Arterial lesions were
126 defined by the presence of aortic or peripheral aneurysm, pseudoaneurysm or arterial
127 thrombosis. The presence of a potential triggering trauma was also investigated for each arterial
128 lesion. Patients who did not require surgery or with pulmonary artery aneurysms were excluded
129 from the analysis. The indications for surgery were: symptomatic arterial thrombosis; aortic
130 aneurysm greater than 5 cm in diameter or growing by more than 5 mm in 6 months, or
131 becoming symptomatic; pseudoaneurysm of more than 2 cm in diameter or growing.
132 Emergency management was limited to ruptured aneurysms or patients with acute ischemia.
133 The radiological diagnosis of aneurysm or pseudoaneurysm was based on arterial Doppler
134 ultrasound combined with computed tomography (CT)-scan. The surgical technique and the
135 material used (prosthetic graft, saphenous autograft, arterial allograft, or stent) were analyzed
136 according to the different anatomical locations. Bypass anastomosis was performed as usual.
137 The choice of the technique used was left to the referee surgeon's discretion. In some cases, a
138 sleeving protection was used, consisting in locating a short prosthetic graft segment around the
139 vascular anastomosis (figures 1 and 2) to reinforce it and to prevent secondary pseudoaneurysm
140 formation. This technique was used when the diagnosis of VBD was confirmed or strongly

141 suspected. In these cases, a sleeve protection was used on each end-to-end anastomosis remote
142 from a bifurcation. For large bypasses (diameter >8 mm), the same graft segment is used for
143 sleeving. For ≤ 8 mm bypasses, a larger (+1mm) segment than the bypass graft is required to
144 avoid stenosis.

145 The postoperative follow-up was based on clinical examination and ultrasound or CT-scan. The
146 primary patency rate was calculated.

147 A recurrence was defined as the reappearance of aneurysm or pseudoaneurysm at the operated
148 site or in another location, or by the occurrence of bypass thrombosis.

149 Early recurrence was defined as the occurrence of arterial aneurysm within less than 1 year,
150 confirmed by arterial Doppler ultrasound or CT-scan. Global doppler ultrasound was
151 systematically performed 6 months after surgery and yearly thereafter during the follow-up. In
152 case of thoracic aortic repair, a CT-scan was performed at the same timepoints. Recurrence at
153 the same arterial site and recurrence at a new site were differentiated. The follow-up was
154 performed by the surgical department and by the referent BD medical department located in the
155 same hospital.

156 The number of recurrences was compared according to the nature of the implanted arterial
157 substitute: synthetic graft, venous autograft, arterial allograft, or stent. The effect of sleeve
158 anastomosis on recurrences was assessed. The recurrence rate in patients receiving dedicated
159 medical treatment for BD was also compared to the recurrence rate in untreated patients,
160 including those who could have benefited from a sleeving or not. Steroids and
161 immunosuppressive treatment were systematically prescribed prior to surgery in BD patients
162 from our institution. However, in some cases, no immunosuppressive treatment was given when
163 patients were referred from other centers or when the diagnosis of BD had not been confirmed
164 prior to surgery.

165

166 Ethical issues

167 The study was conducted in accordance with the ethical principles of the Declaration of
168 Helsinki.

169 The study protocol was assessed by the institutional review board of the *Hôpital Universitaire*
170 *Pitié-Salpêtrière*. Patients were informed that their clinical data were likely to be collected, but
171 in accordance with the French law, since this was an observational study involving only routine
172 clinical practices and therefore with no risk for patients, no informed consent was needed.

173

174 Statistical analysis

175 Statistical analyses were carried out using SAS 9.4 software. Quantitative data with normal
176 distribution are expressed as the mean \pm standard deviation and quantitative data with non-
177 normal distribution as the median (interquartile). The recurrence rate according to the effect of
178 the anastomotic sleeve technique was assessed using the Kaplan-Meier method. The Fischer's
179 exact test was used to study the effect of preoperative and postoperative treatments on the
180 occurrence of recurrences (thrombosis, false aneurysm). A P-value less than 0.05 was
181 considered statistically significant.

182

183 **Results**

184 Population

185 Twenty-three BD patients with arterial involvement were included in our study. A total of 47
186 procedures were performed for arterial lesion, with a mean number of 2.4 ± 0.6 procedures per
187 patient in the 17 patients who experienced recurrence. Except for 7 initial interventions
188 performed in another hospital before the diagnosis of BD, all interventions were performed in
189 our department, including after relapses. Patient demographics are presented in **Table I**. At the

190 time of arterial lesions, the main active clinical sign of BD was oral and genital ulcerations in
191 61% of cases. Peripheral locations were more common than aortic ones (Table II).
192 Among patients with peripheral aneurysms, the femoro-popliteal location (48%) was the most
193 common. Among all patients, the first arterial involvement was fusiform aneurysm (n=10),
194 pseudoaneurysm (n=12), or arterial thrombosis (n=1). During the follow-up, including relapses,
195 the treated lesions were pseudoaneurysms in 29 procedures (62%), fusiform aneurysms in 11
196 procedures (23%) and arterial thrombosis in 7 procedures (15%). The lesions were bilateral in
197 2 patients (4%): one patient had bilateral iliac thrombosis and one patient had spontaneous
198 bilateral femoral pseudoaneurysms. Arterial puncture resulted in pseudoaneurysm in 2 cases
199 (4%). Emergency management for ruptured aortic pseudoaneurysm below the renal arteries was
200 needed in two cases (4%).

201

202 Surgical technique

203 Among the 47 procedures, 27 prosthetic grafts (57%), 6 venous autologous grafts (13%), 5
204 cryopreserved allografts (11%) and 3 stent grafts (6%) were used (Table III). Prosthetic grafts
205 were polyethylene terephthalate (PET) in all cases except one. In this case, graft made with
206 polytetrafluoroethylene (PTFE) was used. All anastomoses were performed using
207 polypropylene suture thread. Stent grafts used were Valiant thoracic endograft (Medtronic,
208 Santa Rosa, CA, USA) in one case and covered stents in two cases (fig 3) for peripheral
209 aneurysms. In 6 cases, reconstruction did not require any material (13%) because short resection
210 with direct end-to-end anastomosis could be performed. The vascular substitutes used,
211 depending on the lesion location, are shown in Table III. A total of 95 anastomoses were
212 performed during the 47 procedures. The sleeving technique was used for 25 anastomoses
213 (corresponding to 13 bypass procedures): 23 (92%) were graft-to-artery anastomosis (8 aortic,
214 3 iliac, 11 femoral, and 1 popliteal) and 2 (8%) were venous-to-femoral artery anastomosis.

215 The mean follow-up duration was 8.4 ± 7.5 years. The postoperative mortality rate was 2%.
216 One patient died 1 month after surgery. This patient underwent aortic stent graft in zone 0 with
217 supra-aortic trunk debranching for false aneurysm of the aortic arch. He experienced a septic
218 rupture of the brachiocephalic trunk anastomosis due to *Staphylococcus aureus* graft infection.
219 The postoperative morbidity rate was 8% (4/47). Two patients experienced hematoma at the
220 surgical site, one patient had acute renal failure without dialysis after aortic surgery, and one
221 patient had acute lower limb ischemia due to embolization after treatment of abdominal aortic
222 aneurysm.
223 The 5-year primary and secondary patency rates were 94 and 100%, respectively.
224 All patients received antiplatelet therapy (160 mg of acetylsalicylic acid) after bypass surgery
225 regardless of the substitute used. In case of thrombosis, anticoagulant treatment (vitamin K
226 antagonist) was initiated instead of antiplatelet therapy.

227

228 Recurrence after vascular repair

229 A total of 24 recurrences were observed in 17 patients, corresponding to a recurrence rate of
230 51%. Eleven patients (48%) had 1 recurrence, 5 patients (22%) had 2 recurrences and 1 patient
231 (4%) had 3 recurrences. In 22 cases (92%), the same arterial anatomical site was involved. Only
232 two patients experienced a recurrence in a different site: one patient had contralateral common
233 femoral artery pseudoaneurysm and one had aortic dilation above a previous infra-renal aortic
234 repair.
235 Lesions recurred as anastomotic pseudoaneurysm in 71% of cases (17/24), bypass thrombosis
236 in 25% of cases (6/24) and aortic fusiform aneurysm above previous infra-renal aortic aneurysm
237 repair in 4% of cases (1/24). Graft thrombosis occurred in three patients (femoro-femoral
238 bypass in two cases and aorto-aortic graft in one case). Graft thrombosis recurred one, two and
239 three times respectively in these three patients.

240 The mean time to recurrence for the overall population was 65 ± 68 months. Among the 24
241 recurrences, 7 (24%) occurred within one year. In the 16 cases that relapsed after one year, the
242 mean time to recurrence was 79.6 ± 59 months. In this group, the median time between the first
243 arterial repair and the first recurrence was 45 months (1-232) and it was 18 months (6-143)
244 between the second intervention and the second relapse. In one patient, the time between the
245 third intervention and the third relapse was 20 years. Recurrences according to the time are
246 presented in **Figure 3**.

247 Anastomotic pseudoaneurysm occurred within the first year in 41% of cases (7/17).

248 The nature of the recurrence observed according to the type of arterial repair is shown in **Table**
249 **IV**. The differences highlighted were not statistically significant due to the small size of the
250 sample.

251 In 45 out of the 47 procedures performed, the use of pre- or postoperative immunosuppressive
252 treatment was known. Medical treatment included colchicine, steroids or immunosuppressants.
253 Patients received medical treatment preoperatively in 55% of cases (25/45), postoperatively in
254 89% of cases (40/45) and both pre- and postoperatively in 51% of cases (23/45). No medical
255 treatment was given in 11% (5/45) of cases. Perioperative medical treatments are detailed in
256 **Table V**.

257 The effect of preoperative immunosuppressive treatment on the recurrence rate showed that
258 28% of procedures (7/25) were followed by a recurrence in treated patients compared to 75%
259 (15/20) in untreated patients (OR: 7.31; 95% CI (1.73; 36.81)) (P = 0.002).

260 The effect of postoperative immunosuppressive treatment was also assessed. Forty-two percent
261 of patients who received postoperative treatment (17/40) experienced a recurrence *versus* 80%
262 of untreated patients (4/5) (OR: 5.22; 95% CI (0.46; 277.32)) (P = 0.16).

263 All patients who did not receive any medication before or after surgery (5/5) experienced a
264 recurrence.

265 The effect of the sleeving technique on the occurrence of pseudoaneurysms at anastomosis sites
266 was analyzed and each anastomosis was considered according to the use of a sleeve protection
267 or not. A total of 95 anastomoses were performed, of which 25 were sheathed by a sleeve. The
268 overall pseudoaneurysm rate was 18% (17/95 anastomoses). In the absence of sleeve, the false
269 aneurysm rate was 21% (16/70) but it was only 8% (2/25) in the presence of a sleeve (OR: 2.44;
270 95% CI (0.89; 6.73)) (p = 0.08) (Figure 4).

271 Regarding patients who received pre- and postoperative medical treatments without sleeve, the
272 recurrence rate for each anastomosis was 32% (12/38 anastomoses), while patients who
273 received medical treatments and sleeve showed a recurrence rate of only 10% (2/19
274 anastomoses) (OR: 0.26; 95% CI (0.03; 1.4)) (p = 0.108).

275

276

277 **Discussion**

278 We provide here the results of a large series of patients who underwent a vascular procedure
279 for arterial lesions of BD. The postoperative mortality rate was low but we observed a high rate
280 of relapse following surgical procedures comparable to those previously published, although
281 our follow-up of 8.4 years was longer¹⁸⁻²⁴. In our series, 51% of BD patients operated for arterial
282 lesions (aneurysms or thrombosis) experienced a relapse which occurred during the first year
283 following surgery in one third of cases.

284 In previous series, recurrence rates of 50% and 56% and postoperative mortality rates of 14%
285 and 15% have respectively been reported in 32 patients who underwent bypass²⁵⁻²⁹ and in 48
286 patients who underwent graft interposition^{22, 26, 28-30}.

287 Device

288 Vasculitis in BD is distinguishable because both arteries and veins of all sizes are involved.
289 Venous complications are more common than arterial lesions in BD, occurring in 30% of

290 patients⁵⁻⁷. VBD patients are at risk of multiple vessel-related complications^{9, 27, 31}. Because
291 of the risk of concomitant venous failure due to BD, we did not use the saphenous vein except
292 in specific cases such as distal bypass or septic surgery. In this study, we used saphenous vein
293 grafts in only 13% of cases and the complication rate was of 50%, exclusively due to
294 pseudoaneurysm formation. In fact, results did not significantly differ from those of prosthetic
295 grafts, but these findings should be interpreted with caution because of the small number of
296 venous bypass procedures performed.

297 The use of prosthetic grafts was associated with a relapse rate of 43%. Thrombosis and
298 pseudoaneurysm occurred after 4/27 (14.8%) and 9/27 (33.3%) procedures, respectively.
299 Thrombosis could be explained by the hypercoagulability secondary to inflammation due to
300 BD in these young patients without atheroma³².

301 Arterial allograft could be another option as we observed a lower complication rate of 20%, but
302 in a limited number of cases (1/5 cases).

303 Arterial wall injury secondary to anastomosis performed between a native artery and a vein or
304 prosthetic graft is a local risk factor for relapse. The endovascular approach has recently been
305 shown to improve results¹⁸ although there may be trauma between a stent and the vascular wall.
306 The endovascular approach was used in only 6% of cases in our study and did not allow
307 concluding to any difference. The endovascular approach could be effective because no
308 anastomosis is needed. The stent should be long enough to cover the lesion in order to seal in a
309 healthy part of the vessel³³. However, arterial puncture is a risk factor for pseudoaneurysm
310 formation^{23, 34, 35} (3 cases in our study). Moreover, data on the long-term results are limited in
311 the literature³⁶.

312 Endovascular procedures are also increasingly used in the management of BD, but their
313 superiority has not been demonstrated. Except for Balcioglu et al³⁷ who have not observed any
314 aneurysm formation nor thrombosis after aortic stent graft placement after a median follow-up

315 of 40 months, all other series have reported a recurrence rate ranging between 8.3 and 75% after
316 endovascular procedures^{18-21, 29, 34, 38-40}. Many cases of pseudoaneurysms at the proximal or
317 distal margin of the stent graft have been described^{21, 35, 40, 41} and resulted in patient death in
318 three cases due to rupture^{19, 29, 40}. Despite the less invasive nature of endovascular treatment,
319 interactions between the stentgraft and the vascular wall led to the formation of
320 pseudoaneurysms at both ends. Yin et al¹⁸ have shown that limiting the oversizing to less than
321 5% could decrease this risk.

322 Liu et al²⁰ have shown better results with bypass surgery than with stent graft in 18 procedures
323 on peripheral arteries. However, the results of aortic aneurysm exclusion were disappointing:
324 among 22 procedures for aortic pseudoaneurysms, a stent graft was used in 19 cases, resulting
325 in recurrence and mortality rates of 18% and 16%, respectively, after a short median follow-up
326 of 23 months. Kim et al²³ have reported the experience of 20 aneurysms in 16 patients treated
327 with stent grafts after a mean follow-up of 4 years. The relapse rate of 19% was low compared
328 to 8 aneurysms in 7 patients who underwent open repair with a relapse rate of 43%.

329 However, the technique consisting in performing bypass without anastomosis, as described in
330 the Viabahn Padova sutureless technique, could be an option⁴².

331

332 **Relapses**

333 In the present study, the recurrence involved the same arterial anatomical site in 92% of cases.
334 Patients who received perioperative steroids and/or immunosuppressants showed a reduced risk
335 of relapse. In line with previous studies and the European League Against Rheumatism
336 (EULAR) recommendations^{20, 26, 27, 31, 36, 37, 43}, anastomotic relapses and graft thrombosis in this
337 series were less common in patients who received aggressive perioperative medical treatment
338 (corticosteroids and immunosuppressants).

339 In our series, patients were treated preoperatively in only 45% of cases but in these cases, the
340 recurrence rate was significantly reduced compared to that found in untreated patients (28%
341 *versus* 75%). In most cases, BD was not diagnosed at the time of surgery.

342 The use of postoperative treatments effectively decreased the recurrence rate of arterial lesions.
343 Many recurrences (24%) occurred during the first year following the surgical procedure but
344 most of them (76%) were delayed with a mean time to recurrence of 6.5 years. These results
345 showed that the risk of recurrence was sustained, whereas in the literature the mean time to
346 recurrence is 10.5 months after a mean follow-up of 6 years^{24, 31, 32, 39}. However, there is
347 currently no consensus regarding the postoperative follow-up of these patients. Therefore,
348 extended monitoring with repeated Doppler ultrasound explorations could be useful to detect
349 as soon as possible early and delayed local recurrences or monitoring with CT-scan for thoracic
350 aorta and intrathoracic supra-aortic trunks.

351

352 Sleeve anastomosis

353 We showed that recurrences occurred mainly at the same site, especially anastomotic
354 pseudoaneurysms, even when optimal medical treatment was given. That is why we added a
355 surgical technique to decrease this risk. Sleeve protection of the anastomosis is an easy and
356 reproducible technique consisting in reinforcing arterial anastomosis by wrapping it with a
357 larger prosthetic graft. This technique had previously been described^{34, 44} but its effects on the
358 risk of recurrence had never been specifically investigated. In our series, we found that
359 anastomoses protected by a sleeve were less often affected by false aneurysm during the follow-
360 up than those that were not protected. This extra-arterial reinforcement provides a mechanical
361 support that decreases the risk of secondary anastomotic rupture, although this result was not
362 statistically significant and further studies are needed to confirm it.

363 This technique was not systematically used when the diagnosis of BD was not suspected at the
364 time of the first surgical procedure or when anastomosis was too close to a collateral artery
365 (such as renal artery or profundis femoral artery). After a mean follow-up of 8 years, it
366 appeared that the rate of anastomotic false aneurysms was decreased when a sleeve was used.
367 Furthermore, even in patients who received medical treatment, adding a sleeve also decreased
368 the risk of anastomotic false aneurysm. The recurrence rate dropped from 32% to 10% of all
369 anastomoses when a sleeve was associated with the surgical procedure.

370

371 The main limitation of this study is its retrospective design. This disease is not common and
372 the diagnosis is often made after the occurrence of one or two arterial events, which limits the
373 possibility of a prospective approach. Moreover, given the small number of patients, the risk of
374 a type 2 statistical error cannot be ruled out. For this reason, we can only conclude that
375 complications tended to decrease when immunosuppressive treatment and the sleeve protection
376 technique were used.

377

378 **Conclusion**

379 Relapse is a major concern after surgical repair of arterial BD. However, the risk seems to be
380 reduced when medical treatment is used in combination with a vascular procedure. This study
381 suggests the need for targeted perioperative medical management to reduce the risk of arterial
382 recurrence in BD patients. To this end, a multidisciplinary approach combining rheumatologists
383 and vascular surgeons is mandatory.

384 The use of sleeve anastomosis may reduce the risk of recurrence in the form of anastomotic
385 pseudoaneurysms. However, further studies are needed to confirm its efficacy.

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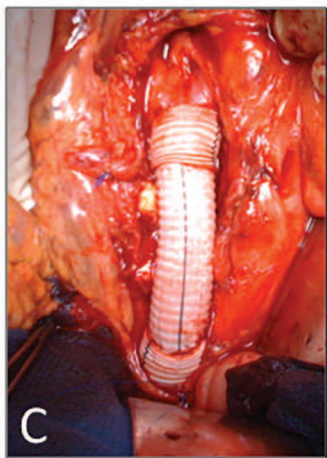
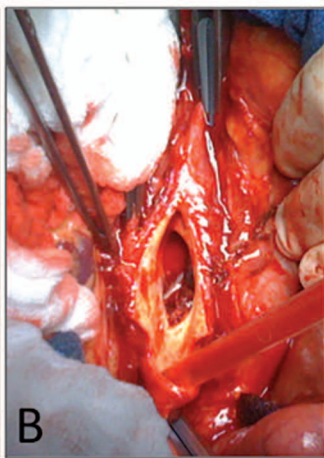
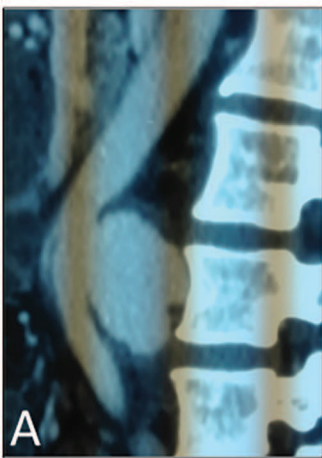
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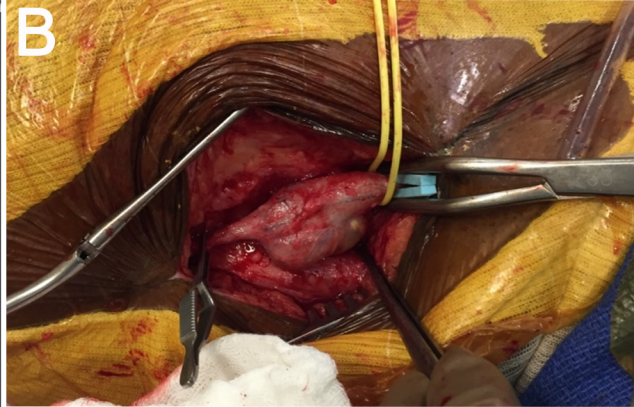
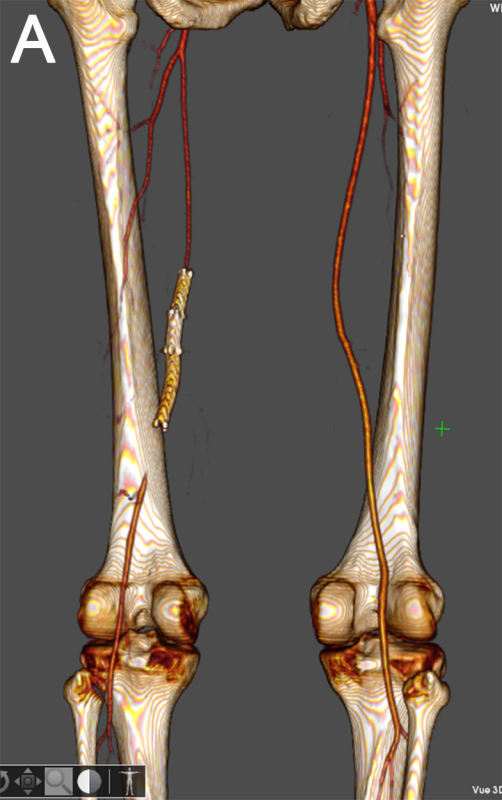
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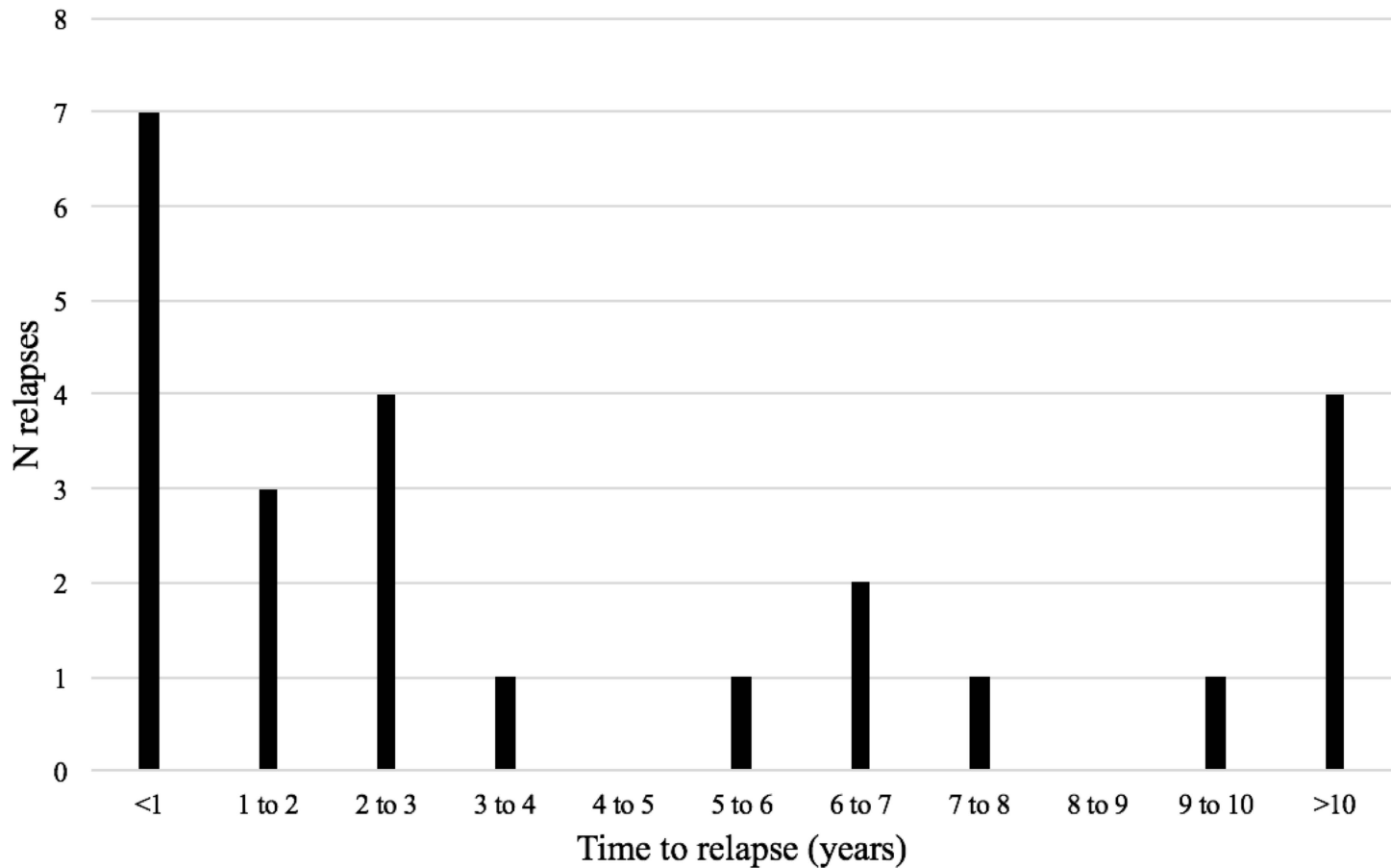
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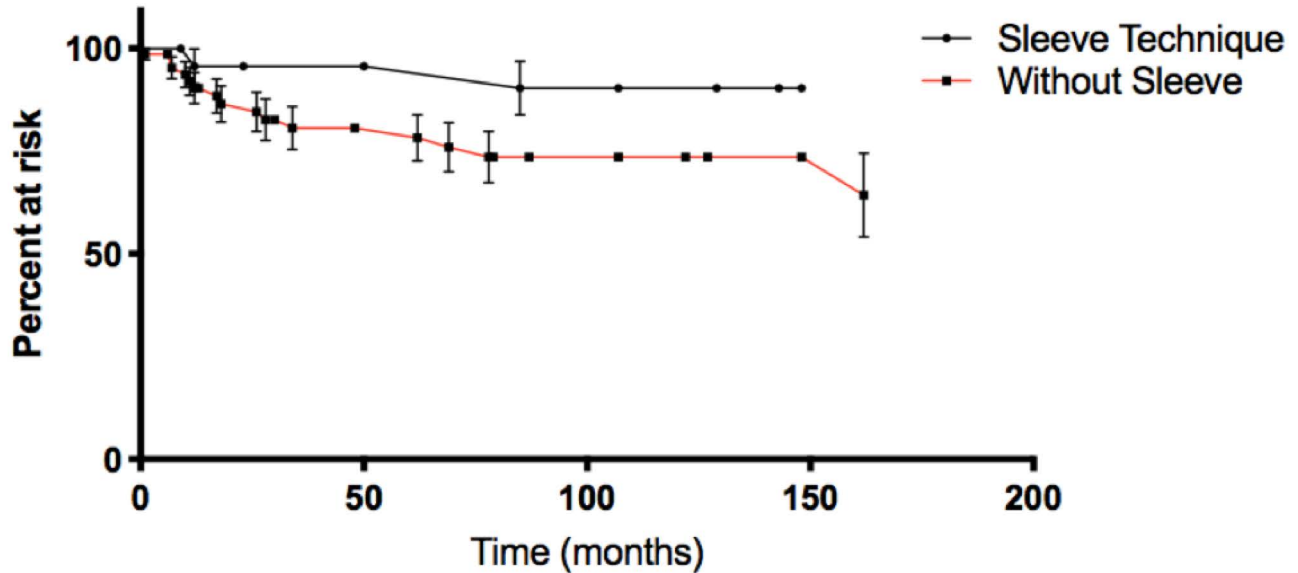




Relapses



Relapse



Time (months)	0	7	10	12	23	50	85	107	129	148	162
Sleeve Technique	25			23	22	20	18	17	13	7	
S.E. (%)				4.2	4.2	4.3	6.5	6.5	6.5	6.5	
Without Sleeve	70	60	58	56	46	38	31	23	18	11	8
S.E. (%)		2.6	3	3.7	4.5	5.6	6.5	6.5	6.7	8.1	9.7

Legends of figures

Figure 1

Spontaneous infrarenal aortic false aneurysm in a patient with BD. (A): Sagittal view of a posterior false aneurysm emerging from the abdominal aorta on CT-scan. (B): Typical aspect of arterial wall defect in the posterior wall of the aorta (after anterior aortotomy). (C): Aorto-aortic bypass with proximal and distal sleeve of the anastomosis with two short segments of a larger graft.

Figure 2

Thrombosis of femoral stent graft after exclusion of superficial femoral artery false aneurysm treated with femoro-femoral prosthetic bypass with sleeve anastomosis. Thrombosis occurred 3 months after stent placement while the patient was treated with 160 mg of aspirin. (A): Lower limb CT-scan showing superficial femoral artery occlusion. (B): Peroperative view of false aneurysm surrounding the stent graft. (C): Femoro-femoral prosthetic bypass with proximal and distal sleeve anastomosis.

Figure 3

Number of recurrences (false aneurysm or thrombosis) according to time after surgery.

Figure 4

Long-term recurrence rate of false aneurysms according to the use of the sleeve technique compared to no sleeve (figures at risk are above the horizontal axis).

Tables

Table I: Demographics, symptoms associated with Behçet's disease at the time of diagnosis of arterial lesions.

Demographics n=23 patients	
Male	17 (74)
Female	6 (26)
Age (years)	34±13
Behçet's disease criteria	
Oral aphthosis	14 (61)
Genital aphthosis	14 (61)
Ocular lesions	2 (9)
Skin lesions	6 (26)
Neurological manifestations	1 (4)
Vascular manifestations	23 (100) (9)
Cardiovascular risk factors	
High blood pressure	7 (30)
Diabetes	1 (4)
Smoking	5 (22)
Dyslipidemia	2 (9)

Data are expressed as n, % and mean±SD

Table II: Type and location of arterial lesions at the time of diagnosis and after recurrence.

Lesion location	Initial lesion (n=23) N (%)	Including recurrence (n=47) N (%)
Aorta	11 (48)	19 (40)
Thoracic	3	5
Abdominal	8	14
Peripheral	12 (52)	28 (60)
Supra-aortic trunk	1	1
Iliac	2	3
Femoro-popliteal	8	22
Upper limb	1	2
Type of lesion		
Fusiform aneurysm	10 (44)	11 (23)
Pseudoaneurysm	12 (52)	29 (62)
Thrombosis	1 (4)	7 (15)

Table III: Vascular substitutes used according to arterial lesion location.

Arterial lesion location (N=47)	Vascular device				
	Saphenous vein	Graft	Allograft	Stent graft	None
Aortic	0	14	4	1	0
Thoracic	0	3	1	1	0
Abdominal	0	11	3	0	0
Peripheral	6	13	1	2	6
Supra-aortic trunks	0	1	0	0	0
Iliac	0	3	0	0	0
Femoro-popliteal	5	9	1	2	5
Upper limb	1	0	0	0	1

Table IV: Recurrences according to the vascular substitute used.

Recurrence	Vein (N=6)	Graft (N=27)	Allograft (N=5)	Stent (N=3)	None (N=6)
False aneurysm	3	9	1	2	3
True aneurysm	0	1*	0	0	0
Thrombosis	0	4	0	1	1
Recurrence rate (%)	50	43	20	100	67

- One true aneurysm occurred above the proximal anastomosis of an aorto-aortic graft.

Table V: Medical treatment combined with surgery.

Treatment (N=47)	Preoperative (n)	Postoperative (n)	Preoperative + Postoperative (n)
None	20	5	5
Colchicine alone	9	9	5
Corticosteroids and/or immunosuppressants	16	31	18