Arteriovenous fistulas (AVF) for clotting factor administration in adults with severe bleeding disorders

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Background: Intravenous self-infusion of clotting factor concentrates is difficult in adult hemophilia patients with poorly accessible peripheral veins or with significant elbow arthropathy. In such cases AVF are alternatives to central venous devices. They provide ease of access with minimal requirements for care. There have been two reports of successful creation and use of AVF in children with hemophilia, together totaling 53 AVF procedures in 47 patients, all but one of whom were aged ≤ 13[1,2]. We previously reported our experience with AVF in 5 adult hemophilia patients. We now present an updated report of 7 patients.

[1]McCarthy et al. Journal of Vasc Surg 2007; 45:986-990 [2]Mancuso et al 2009. Haematologica; 94:687-692

Methods: Anatomy was suitable for AVF creation in seven of eight potential candidates. Four were on continuous or intermittent prophylactic infusion schedules and three were treated on-demand. Their ages were 18–72 years at the time of AVF creation (median 40). Two have severe hemophilia A (HA) with histories of high titer inhibitors that were fully or partially tolerized; three have moderately severe HA; one has severe hemophilia B (HB); one has type 3 VWD. In one patient the AVF had been created elsewhere in 1995 for hemodialysis, and has been used for factor VIII infusions since he received a renal allograft. The other six AVF were created in our institution between November 2013 and October 2015, four of them by the same vascular surgeon. Of these six, three AVF were radio-cephalic, two were brachiocephalic, and one was ulnar-cephalic.

Results: Patient details are shown in the Table. There was no major surgical blood loss or other complications; local edema was noted in one patient. Clotting factor usage perioperatively (4–6 days) was 152–335 IU/kg. All AVF matured and are being accessed for home self-infusion. Two patients required angioplasty for stenosis post-operatively, one at 2 months and one at 6 months. One patient remains on an on-demand treatment schedule; the others are now on prophylactic schedules, either alternate day or twice weekly (the HB patient).

Patient	Diagnosis	Age at AVF	Regimen pre-AVF	Regimen post-AVF	Compliance pre-AVF	Compliance post-AVF	Surgical details	Complications
1	Moderate HA (3%)	N/A	On-demand	On-demand	Good	Good	N/A	N/A
2	VWD type 3	18	Prophylaxis alternate day	Prophylaxis alternate day	Good	Good	Left radio-cephalic	None
3	Moderate HA (3%)	72	Prophylaxis alternate day	Prophylaxis alternate day	Fair	Good	Right radio-cephalic	None
4	Moderate HA (2%)	33	On-demand (failed prophylaxis)	Prophylaxis alternate day	Fair	Fair	Left ulnar-cephalic	None
5	Severe HB (<1%)	46	Intermittent prophylaxis	Prophylaxis twice weekly		Good	Right radio-cephalic	Angioplasty 6 months
6	Severe HA with inhibitor	22	ITI with daily VWF/FVIII	ITI with daily VWF/FVIII	Good	Good	Left brachio-cephalic	None
7	Severe HA (tolerized inhibitor)	69	Prophylaxis alternate day	Prophylaxis alternate day	Good	Good	Right brachiocephalic	Angioplasty 2 months

Conclusion: AVF can facilitate venous access for home management of hemophilia in adults who have difficulty accessing native veins.