

**On-line Table 1: Clinical data on patients with PMAVs at presentation and long-term follow-up**

| No. | M/F | Age (yr) | Site   | Type | TTDx (mo) | TTRx (mo) | Symptoms               | Pre Amb ALS | Pre Mict ALS | Treatment       | Post Amb ALS | Post Mict ALS | FU Angio | FU (mo) |
|-----|-----|----------|--------|------|-----------|-----------|------------------------|-------------|--------------|-----------------|--------------|---------------|----------|---------|
| 1   | F   | 2        | T10    | 3    | —         | —         | Progressive myelo      | 4           | 0            | Embo only       | 1            | 0             | Cure     | 112     |
| 2   | F   | 6        | T8–9   | 3    | 0.5       | 0.5       | Acute myelo, SAH       | 1           | 0            | Embo only       | 0            | 0             | Cure     | 4       |
| 3   | F   | 7        | L2     | 3    | —         | —         | SAH                    | 4           | 0            | Embo × 2        | 0            | 0             | Cure     | 41      |
| 4   | F   | 7        | T1     | 3    | 0.07      | 0.07      | Acute para, SAH        | 5           | 3            | Embo × 2, OR    | 1            | 2             | Fistula  | 128     |
| 5   | F   | 11       | C4     | 3    | 0         | 1         | SAH                    | 0           | 0            | Embo × 2, OR    | 0            | 0             | Cure     | 36      |
| 6   | F   | 12       | T6     | 3    | 0.75      | 0.75      | Acute para, EDH        | 5           | 3            | Embo × 4, OR    | 5            | 3             | Fistula  | 24      |
| 7   | M   | 13       | L1–3   | 3    | —         | —         | SAH, hemato            | 5           | 3            | Embo, OR        | 0            | 0             | Cure     | 4       |
| 8   | M   | 16       | T12–L1 | 2    | 0.23      | —         | Acute myelo            | 2           | 3            | OR              | —            | —             | NA       | —       |
| 9   | M   | 22       | T10    | 3    | —         | —         | Recurrent SAH          | 3           | 2            | Embo, OR        | 2            | 2             | NA       | 84      |
| 10  | F   | 23       | L1     | 2    | —         | —         | Acute myelo, hemato    | 4           | 2            | Embo, OR        | 3            | 2             | Cure     | 1       |
| 11  | F   | 24       | L2     | 2    | —         | —         | Progressive myelo, SAH | 4           | 3            | OR × 2/Embo × 3 | 2            | 2             | Cure     | 228     |
| 12  | M   | 27       | T12–L1 | 2    | 24        | 24        | Progressive myelo      | 4           | 3            | OR × 2          | 3            | 2             | Cure     | 1       |
| 13  | M   | 31       | L1     | 3    | 60        | 60        | Progressive myelo      | 4           | 3            | Embo × 2/OR     | 3            | 3             | Cure     | 108     |
| 14  | M   | 31       | L1     | 2    | 10        | 10        | Progressive myelo      | 2           | 1            | Embo × 1/OR × 2 | 3            | 1             | Fistula  | 7       |
| 15  | F   | 32       | T12    | 2    | 36        | 36        | Progressive myelo      | 4           | 2            | Embo × 2/OR     | 1            | 1             | Cure     | 63      |
| 16  | M   | 36       | T7–9   | 3    | 96        | 96        | Progressive myelo      | 3           | 1            | Embo, OR        | 2            | 3             | Cure     | 84      |
| 17  | F   | 39       | L2     | 2    | 4         | 4         | Progressive myelo      | 4           | 2            | Embo × 1/OR × 2 | 3            | 2             | Cure     | 22      |
| 18  | F   | 40       | T10–11 | 3    | 3         | 3         | Progressive myelo      | 3           | 0            | Embo × 2/OR     | 0            | 1             | Cure     | 54      |
| 19  | M   | 40       | T10    | 1    | 48        | 120       | Progressive myelo      | 2           | 0            | OR              | 1            | 0             | Cure     | 120     |
| 20  | F   | 42       | T11–12 | 2    | —         | —         | Progressive myelo      | 4           | 2            | Embo × 3/OR × 3 | 3            | 3             | Cure     | 108     |
| 21  | M   | 43       | T11    | 1    | 0.1       | 0.1       | Acute para, SAH        | 5           | 3            | OR × 2          | 4            | 1             | Cure     | 2.5     |
| 22  | F   | 44       | T11    | 2    | 0.13      | 0.13      | SAH                    | 2           | 0            | OR              | 1            | 0             | Cure     | 171     |
| 23  | F   | 52       | L1     | 2    | 6         | 7         | Progressive myelo      | 2           | 0            | OR              | 1            | 0             | Cure     | 1       |
| 24  | F   | 55       | L2     | 2    | 6         | 6         | Progressive myelo      | 2           | 3            | OR              | 2            | 3             | Cure     | 22      |
| 25  | M   | 57       | L2     | 1    | 12        | 12        | Progressive myelo      | 5           | 3            | OR              | 4            | 3             | Cure     | 2       |
| 26  | M   | 58       | T12    | 1    | 18        | 18        | Progressive myelo      | 2           | 0            | OR              | 3            | 1             | Fistula  | 45      |
| 27  | M   | 59       | C1–2   | 1    | 1         | 1         | Acute myelo            | 4           | 2            | Embo, OR        | 3            | 0             | Cure     | 3       |
| 28  | F   | 65       | L1     | 1    | 0.25      | —         | Acute para             | 5           | 3            | None            | 2            | 3             | NA       | 60      |
| 29  | F   | 68       | L2     | 2    | 12        | 12        | Progressive myelo      | 2           | 2            | OR              | 0            | 1             | Cure     | 2.5     |
| 30  | M   | 71       | T9–11  | 2    | 6         | —         | Progressive myelo      | 2           | 1            | None            | —            | —             | NA       | —       |
| 31  | F   | 71       | T11    | 2    | 16        | 16        | Progressive myelo      | 2           | 3            | OR × 2          | 2            | 3             | Fistula  | 68      |
| 32  | F   | 82       | T9     | 2    | 36        | 36        | Progressive myelo      | 4           | 1            | Embo only       | 4            | 1             | Fistula  | 6       |

**On-line Table 2: Clinical presentation**

| Chronic (n = 20)       |               |              | Acute (n = 12) |        |            |
|------------------------|---------------|--------------|----------------|--------|------------|
| 19                     | 1             | 7            | 1              | 1      | 3          |
| Progressive myelopathy | Recurrent SAH | SAH          | EDH            | Hemato | Acute para |
| 1                      |               | 1            |                |        |            |
| Acute SAH              |               | Hematomyelia |                |        |            |

**On-line Table 3: Outcome based on angiographic cure versus residual fistula**

| Fistula              | Age <sup>a</sup> | Presentation            | Changes in Ambulation |             |
|----------------------|------------------|-------------------------|-----------------------|-------------|
|                      |                  |                         | No Change/Worse       | Improvement |
| Residual (n = 6)     | 44 (7–82)        | Acute = 2, chronic = 4  | 5                     | 1           |
| No residual (n = 22) | 34 (2–68)        | Acute = 8, chronic = 14 | 2                     | 20          |

<sup>a</sup> Age expressed in mean years (range).

**On-line Table 4: Outcome based on angiographic cure versus residual fistula**

| Fistula                      | Age <sup>a</sup> | Presentation            | Changes in Micturition |             |
|------------------------------|------------------|-------------------------|------------------------|-------------|
|                              |                  |                         | No Change/Worse        | Improvement |
| Residual ( <i>n</i> = 6)     | 44 (7–82)        | Acute = 2, chronic = 4  | 5                      | 1           |
| No residual ( <i>n</i> = 22) | 34 (2–68)        | Acute = 8, chronic = 14 | 15                     | 7           |

<sup>a</sup> Age expressed in mean years (range).

**On-line Table 5: Type of fistula**

|                        | 1 ( <i>n</i> = 6) | 2 ( <i>n</i> = 15) | 3 ( <i>n</i> = 11) |
|------------------------|-------------------|--------------------|--------------------|
| Age (range) (yr)       | 54 (40–65)        | 45 (16–82)         | 17 (2–40)          |
| Sex                    |                   |                    |                    |
| Male                   | 5                 | 4                  | 4                  |
| Female                 | 1                 | 11                 | 7                  |
| Location               |                   |                    |                    |
| Cervical               | 1                 | 0                  | 1                  |
| Thoracic               | 3                 | 6                  | 7                  |
| Thoracolumbar junction | 0                 | 2                  | 0                  |
| Lumbar                 | 2                 | 7                  | 3                  |
| Symptoms               |                   |                    |                    |
| Progressive myelo      | 3                 | 12                 | 4                  |
| Acute myelo            | 1                 | 2                  | 1                  |
| Acute para             | 2                 | 0                  | 2                  |
| SAH only               | 0                 | 1                  | 4                  |
| Hemorrhage             | 1                 | 3                  | 7                  |
| TTDx (mo)              | 13.2 (0.1–48)     | 12.1 (0.13–36)     | 22.9 (0–96)        |
| TTRx (mo)              | 30.2 (0.1–120)    | 13.9 (0.13–36)     | 23.1 (0.07–96)     |
| Pre total ALS          | 5.83              | 4.80               | 4.72               |
| Post total ALS         | 4.5               | 3.77               | 2.54               |
| % Change total         | 23% reduction     | 21% reduction      | 46% reduction      |
| Pre amb ALS            | 3.83              | 2.93               | 3.36               |
| Post amb ALS           | 2.83              | 2.15               | 1.27               |
| Change amb             | 26% reduction     | 27% reduction      | 62% reduction      |
| Pre mict ALS           | 2.00              | 1.87               | 1.36               |
| Post mict ALS          | 1.67              | 1.62               | 1.27               |
| Change mict            | 17% reduction     | 13% reduction      | 7% reduction       |
| Treatment              |                   |                    |                    |
| OR                     | 4                 | 7                  | 0                  |
| Embo                   | 0                 | 1                  | 3                  |
| Combo                  | 1                 | 6                  | 8                  |
| NA                     | 1                 | 1                  | 0                  |
| Cure/residual          |                   |                    |                    |
| Cure                   | 4                 | 10                 | 8                  |
| Residual               | 1                 | 3                  | 2                  |
| NA                     | 1                 | 2                  | 1                  |
| FU (range) (yr)        | 38.8 (2–120)      | 53.9 (1–228)       | 61.8 (4–128)       |

**On-line Table 6: PMAVF classification by Anson and Spetzler**

|        | Size         | Arterial Feeder  | Fistula Flow          | Venous System                       |
|--------|--------------|------------------|-----------------------|-------------------------------------|
| Type A | Small        | Single branch    | Slow                  | Minimal dilation                    |
| Type B | Intermediate | Multiple dilated | Increased flow        | Dilated veins                       |
| Type C | Giant        | Multipediculated | Large high-flow shunt | Markedly dilated and tortuous veins |

**On-line Table 7: PMAVF classification by Merland et al**

|        | Feeding Arteries           | Fistula Flow | Venous Drainage                  |
|--------|----------------------------|--------------|----------------------------------|
| Type 1 | Single ASA                 | Slow         | Mild dilation                    |
| Type 2 | ASA, $\geq 1$ PLSA         | High         | Dilated, tortuous, slow drainage |
| Type 3 | Multiple dilated ASA, PLSA | High         | Venous ectasia, high flow        |

**On-line Table 8: Literature review of PMAVF and outcome**

| No. | Author                      | Year | No. Pts | M/F  | Age (range) <sup>a</sup>                         | Classification                                                                                                                                      | Symptom Duration                                     | Rx                                                                                                                   | FU (range)  | Imaging Outcome                                      | Clinical Outcome                                                                  | Comments                                                                                                                                                                                                                                                                                                                                               |
|-----|-----------------------------|------|---------|------|--------------------------------------------------|-----------------------------------------------------------------------------------------------------------------------------------------------------|------------------------------------------------------|----------------------------------------------------------------------------------------------------------------------|-------------|------------------------------------------------------|-----------------------------------------------------------------------------------|--------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| 1   | Gueguen et al <sup>12</sup> | 1987 | 11      | 5/6  | Onset: 22                                        | Gueguen (similar to Merland)                                                                                                                        | 12 yr                                                | 4 OR                                                                                                                 | 19          |                                                      | 2 much improved                                                                   | 3 SAH; type of fistula did not affect severity of symptoms or prognosis; Rx choice did not affect outcome; time to treatment affected outcome                                                                                                                                                                                                          |
| 2   | Mourier et al <sup>7</sup>  | 1993 | 35      | NA   | Dx: 34<br>25 (2–42)                              | Merland:<br>4 type 1,<br>9 type 2,<br>22 type 3                                                                                                     | 9 yr<br>(5 mo–26 yr)                                 | 5 Embo<br>2 Combo<br><br>6 OR,<br><br>25 Embo<br>(22 balloon,<br>1 glue,<br>2 particles)<br>3 combo,<br>or untreated | 1–20        | 27 cured (angio)                                     | 22 better<br>9 stable<br>2 worse<br><br>4 mildly improved<br>5 stable             | 7 SAH; clinical grade: 1—minimal disability<br>2—comparable with professional life<br>3—severe disability<br>4—confined to bed; all type 3 treated with balloons; persistent bladder/bowel sx post-treatment; cure of type 3 leads to improvement; cure of types 1 and 2 shows 50/50 chance of improvement<br>Worsening sx due to arachnoiditis postop |
| 3   | Halbach et al <sup>8</sup>  | 1993 | 10      | 3/7  | 20 (2–40)                                        | Merland:<br>2 type 2,<br>8 type 3,<br>Halbach:<br>single hole, single feeder,<br>single hole, multiple feeders,<br>multiple holes, multiple feeders |                                                      | 3 Embo<br>7 Combo                                                                                                    | 45          | 7 cured (angio)                                      | 4 dramatic improvement<br>4 modest improvement<br>1 stable<br>1 worse             |                                                                                                                                                                                                                                                                                                                                                        |
| 4   | Barrow et al <sup>19</sup>  | 1994 | 8       | 4/4  | 33 (0–55)                                        | Anson and Spetzler:<br>4 type A,<br>3 type B,<br>1 type C                                                                                           |                                                      | 7 OR with<br>intraop<br>angio,                                                                                       | 1 Embo      |                                                      | 6 better<br>1 stable<br>1 worse                                                   |                                                                                                                                                                                                                                                                                                                                                        |
| 5   | Ricolfi et al <sup>13</sup> | 1997 | 12      | 8/4  | Dx: 12 (0.3–39)<br>Rx: 17 (4–43)                 | Merland: all type 3                                                                                                                                 |                                                      | 12 Embo<br>(particles,<br>balloon)                                                                                   |             | 10 cured (angio)                                     | 6 good<br><br>1 fair<br>3 poor<br>6 better<br>3 stable<br>1 worse                 | 5 SAH; Cobb syndrome; HHT: TFRx = 5 yr (1 week–23 yr); good = independent ambulation; fair = crutches/cane, occasional urinary incontinence/retention; poor = unable to stand, incapacitating urinary symptoms                                                                                                                                         |
| 6   | Hida et al <sup>6</sup>     | 1999 | 20      | 8/12 | 37 (3–67)                                        | Anson and Spetzler:<br>4 type A,<br>11 type B,<br>5 type C                                                                                          |                                                      | 11 OR<br><br>9 Combo                                                                                                 | 54 (12–198) | 16 cured (angio)<br><br>4 residual                   | 11 better<br><br>8 stable<br>1 worse                                              | 11 hemorrhage, outcome measure: Neurosurgical Cervical Spine Scale for lower extremity motor function; residual fistula = no improvement; type A and Better outcome than type C<br>8 stable<br>1 worse                                                                                                                                                 |
| 7   | Cho et al <sup>5</sup>      | 2005 | 19      | 12/7 | 28 (6–52)                                        | Anson and Spetzler:<br>9 type A,<br>6 type B,<br>4 type C                                                                                           | 22 Mo (3 d to 17 yr)                                 | 8 OR<br><br>9 Embo<br>2 Combo<br>16 OR                                                                               | 20 (2–55)   | 14 had follow-up MRI or angio (results not reported) | 9 better<br>(6 type A, 1 type B,<br>2 type C)<br>6 stable<br>4 worse<br>15 better | Type A patients more likely to have improvement in symptoms; no differentiation between ambulation and micturition                                                                                                                                                                                                                                     |
| 8   | Rodesch et al <sup>20</sup> | 2005 | 32      | 22/9 | Children: <i>n</i> = 10<br>Adults: <i>n</i> = 22 | Merland:<br>25 type 1 ("microAVF"),<br>7 type 2/3 ("macroAVF")                                                                                      | Acute:<br><i>n</i> = 18<br>Chronic:<br><i>n</i> = 16 |                                                                                                                      | 40          | 10 cured (angio)                                     | 5 residual (angio)                                                                | Outcome scale is Kamofsky Performance Scale: no surgical outcomes reported                                                                                                                                                                                                                                                                             |

<sup>a</sup> Age (range) = mean age and range in years.