### Mean Basilar Artery Diameter Measurement
The following step-by-step procedure was used to measure the mean basilar artery diameter in the present study:

1. The MRA axial images were loaded from the PACS into Med-station (Exprivia, Molfetta, Italy), a commercially available DICOM viewer in use at our hospital.
2. Axial images were magnified, keeping the vertebral-basilar circle in the middle of the image.
3. Image window was set by selecting a rectangular area of approximately 3 cm × 2 cm (6 cm$^2$) in a cerebellar hemisphere, including both cortex and white matter, while pressing the right button of the mouse together with the "Alt" key. In this way, windowing was optimized for a quite restricted range of intensities, and the vessels appeared rather bright.
4. The diameter of the basilar artery lumen was measured with the function "Linear Measure"; when the axial section was oval-shaped, the minimum diameter was chosen to avoid the diameter oversizing caused by oblique sectioning of the vessel.
5. The first measure was performed just above the conjunction of the 2 vertebral arteries; both diameter and section number were recorded. The second measure was obtained just below the division of the artery into the posterior cerebral arteries; again, both diameter and section number were recorded. The third measure was taken at the midpoint between the above-mentioned measures according to the section number of the previous measurements.

All measures were recorded in a data base, and the mean value of the basilar artery diameter was calculated.

### Curved Length and Linear Length Measurement
The following step-by-step procedure was used to measure basilar artery length and tortuosity in the present study:

1. All MRA native axial partitions were loaded into commercially available software (syngo MR Workplace, Siemens, Erlangen, Germany).
2. The function "Inspace Window" was activated, obtaining interactive 3D multiplanar reconstructions.
3. The "Vessel Analysis" menu was opened, and 2 anatomic basilar artery landmarks were identified based on multiplanar and maximum intensity projection 3D reconstructions: the confluence of the vertebral arteries (proximal/caudal basilar artery extremity) and the bifurcation of the basilar artery into the posterior cerebral arteries (distal/rostral basilar artery extremity).
4. After setting the 2 above-mentioned landmarks with the function "Trace," the software automatically provided a curved line following the vessel path according to the different signal intensity of the basilar artery compared with the surrounding parenchymal structures and subarachnoid spaces. The exact correspondence between the line and the vessel course was accurately verified in all space dimensions.
5. Whenever the correspondence between basilar artery and curved line was suboptimal, the operator manually adjusted the course of the line by activating the function "Edit": this function shows several square dots along the curved line that can be manually positioned centrally in the vessel lumen until the curved line follows the vessel along its whole course (additional square dots can be placed if needed).
6. Activating the functions "Measure" and "Curve," the software provided the length (in mm) of the curved line running from the proximal and distal basilar artery extremities (curved length). After recording the measure, all square dots except the proximal and distal extremes were deleted, obtaining a straight line joining the remaining 2 square dots. The software provided the corresponding measure (linear length [in mm]), which was also recorded. The tortuosity index was eventually calculated according to the following formula:

Tortuosity Index = (curved length ÷ linear length) − 1

**On-line Table 1: Basilar artery quantitative MRA findings in patients with FD and control patients**

| | FD | | | Controls | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|
| Demographics | All | M | F | All | M | F | FD vs C | M-FD vs M-C | M-FD vs F-FD | F-FD vs F-C | M-C vs F-C |
| No. of patients | 110 | 40 | 70 | 108 | 40 | 68 | NA | NA | NA | NA | NA |
| Mean age (yr) | 39.4 ± 18.6 | 36.9 ± 21.3 | 40.9 ± 16.9 | 42.0 ± 18.2 | 38.1 ± 19.5 | 44.3 ± 17.1 | $P = .30$ | $P = .79$ | NA | $P = .41$ | NA |
| Mean diameter (mm) | 4.32 ± 0.72 | 4.07 ± 0.55 | 4.16 ± 0.62 | 3.78 ± 0.36 | 3.66 ± 0.36 | 3.78 ± 0.36 | $P < .001^a$ | $P < .001^a$ | $P = .04^a$ | $P < .001^a$ | $P = .10$ |
| Linear length (mm) | 28.34 ± 4.03 | 28.91 ± 4.4 | 28.02 ± 3.83 | 27.18 ± 3.32 | 27.66 ± 3.16 | 26.86 ± 3.41 | $P = .02^a$ | $P = .15$ | $P = .27$ | $P = .06$ | $P = .23$ |
| Curved length (mm) | 31.13 ± 5.38 | 32.72 ± 5.81 | 30.21 ± 4.92 | 29.88 ± 4.79 | 29.83 ± 4.5 | 29.81 ± 5.02 | $P = .07$ | $P = .01^a$ | $P = .02^a$ | $P = .64$ | $P = .98$ |
| Tortuosity index | 0.097 ± 0.038 | 0.133 ± 0.094 | 0.077 ± 0.070 | 0.099 ± 0.113 | 0.077 ± 0.086 | 0.114 ± 0.125 | $P = .86$ | $P = .006^a$ | $P < .001^a$ | $P = .03^b$ | $P = .10$ |

**Note:**—C indicates controls; F = female; M = male; NA, not available.

[a] Significant $P$ values.

[b] Not significant after adjusting for age.

On-line Table 2: Basilar artery findings in MR studies investigating basilar artery ectasia in patients with Fabry disease

| Study | Demographics | FD | | | Controls | | | Non-FD With Stroke | | | MR Technique | Cutoff |
|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | | All | M | F | All | M | F | All | M | F | | |
| Fellgiebel et al 2011[15] | No. of patients | 25 | 10 | 15 | 20 | 9 | 11 | 26 | 9 | 17 | TOF-MRA (MIP), sagittal plane, mean measure (proximal, middle, distal) | 2.98 mm vs patients with stroke: sensitivity, 84%; specificity, 88.5% |
| | Mean age (yr) | 36.5 ± 11.0 | NA | NA | 36.8 ± 10.0 | NA | NA | 39.8 ± 10 | NA | NA | | |
| | Mean BAD | 3.38 ± 0.59 | 3.88 ± 0.64 | 3.08 ± 0.31 | 2.48 ± 0.36 | NA | NA | 2.45 ± 0.5 | 2.7 ± 0.42 | 2.3 ± 2.52 | | |
| Azevedo et al 2012[19] | No. of patients | 12 | 5 | 12 | NA | NA | NA | NA | NA | NA | TOF-MRA (MIP), sagittal plane, mean measure (proximal, middle, distal) | NA |
| | Mean age (yr) | 35.8 ± 12.8 | 27.4 ± 11.5 | 41.7 ± 10.6 | NA | NA | NA | NA | NA | NA | | |
| | Mean BAD | 3.66 ± 0.72 | NA | NA | NA | NA | NA | NA | NA | NA | | |
| Üçeyler et al 2014[16] | No. of patients | 87 | 30 | 57 | 36 | 14 | 22 | 20 | 13 | 7 | TOF-MRA (MIP), sagittal plane, mid-artery diameter | 3.2 mm vs patients with stroke: sensitivity, 87%; specificity, 86% |
| | Mean age (yr) | NA | 40 | 45 | NA | 44 | 36 | NA | 58 | 75 | | |
| | Mean BAD (range) | NA | 3.5 (2.7–4.4) | 3.1 (2–5) | 2.9 (1.2–5.5) | 2.9 (2.3–5.5) | 2.9 (1.2–3.6) | 3.2 (1.3–4.2) | 3.2 (2.6–4.2) | 3.2 (1.3–3.6) | | |
| Politei et al 2014[17] | No. of patients | 70 | 27 | 43 | NA | NA | NA | NA | NA | NA | TOF-MRA (MIP) and T2-weighted images[a] | NA |
| | Mean age (yr) | 32.6 ± 1.8 | 25.5 ± 2.2 | 37.0 ± 2.3 | NA | NA | NA | NA | NA | NA | | |
| | BAD >4.5 mm | NA (42.6%[a]) | NA (55.5%[a]) | NA (34.8%[a]) | NA | NA | NA | NA | NA | NA | | |
| Present study | No. of patients | 110 | 40 | 70 | 108 | 40 | 68 | NA | NA | NA | TOF-MRA, axial plane, mean measure (proximal, middle, distal) | 4.42 mm: sensitivity, 28.18%; specificity, 97.2% |
| | Mean age (yr) | 39.4 ± 18.6 | 36.9 ± 21.3 | 40.9 ± 16.9 | 42.0 ± 18.2 | 38.1 ± 19.5 | 44.3 ± 17.1 | NA | NA | NA | | |
| | Mean BAD | 4.16 ± 0.62 | 4.32 ± 0.72 | 4.07 ± 0.52 | 3.70 ± 0.36 | 3.78 ± 0.36 | 3.66 ± 0.36 | NA | NA | NA | | |

**Note:**—BAD indicates basilar artery diameter; F = female; M = male; NA, not available.
a Smoker criteria also included increased elongation and vessel displacement.

On-line Table 3: Main data of MR studies investigating basilar artery elongation/tortuosity in patients with Fabry disease

| Study | Demographics | FD | | | Controls | | | Non-FD With Stroke | | | MR Technique |
|---|---|---|---|---|---|---|---|---|---|---|---|
| | | All | M | F | All | M | F | All | M | F | |
| Ginsberg et al 2006[13] | No. of patients | 47 | 32 | 15 | NA | NA | NA | NA | NA | NA | TOF-MRA (MIP) and T2-weighted images |
| | Mean age (yr) | NA | NA | NA | NA | NA | NA | NA | NA | NA | |
| | Dolichoectasia (%) | NA (14.9%[a]) | NA | NA | NA | NA | NA | NA | NA | NA | |
| Politei et al 2014[17] | No. of patients | 70 | 27 | 43 | NA | NA | NA | NA | NA | NA | TOF-MRA (MIP) and T2-weighted images |
| | Mean age (yr) | 32.6 ± 1.8 | 25.5 ± 2.2 | 37.0 ± 2.3 | NA | NA | NA | NA | NA | NA | |
| | Dolichoectasia[b] | 42.9% | 55.5% | 34.8% | NA | NA | NA | NA | NA | NA | |
| Present study | No. of patients | 110 | 40 | 70 | 108 | 40 | 68 | NA | NA | NA | TOF-MRA, axial plane |
| | Mean age (yr) | 39.4 ± 18.6 | 36.9 ± 21.3 | 40.9 ± 16.9 | 42.0 ± 18.2 | 38.1 ± 19.5 | 44.3 ± 17.1 | NA | NA | NA | |
| | Linear length (mm) | 28.34 ± 4.03 | 28.91 ± 4.4 | 28.02 ± 3.83 | 27.18 ± 3.32 | 27.66 ± 3.16 | 26.86 ± 3.41 | NA | NA | NA | |
| | Curved length (mm) | 31.13 ± 5.38 | 32.72 ± 5.81 | 30.21 ± 4.92 | 29.88 ± 4.79 | 29.83 ± 4.5 | 29.81 ± 5.02 | NA | NA | NA | |
| | Tortuosity index | 0.097 ± 0.038 | 0.133 ± 0.094 | 0.077 ± 0.070 | 0.099 ± 0.113 | 0.077 ± 0.086 | 0.114 ± 0.125 | NA | NA | NA | |

**Note:**—F indicates female; M = male; NA, not available.
a Qualitative evaluation also included vessel ectasia.
b According to Smoker's criteria.