Special Theme Topic: Japanese Surveillance of Neuroendovascular Therapy in JR-NET/JR-NET2—Part I

Epidemiology of Dural Arteriovenous Fistula in Japan: Analysis of Japanese Registry of Neuroendovascular Therapy (JR-NET2)

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Abstract

We developed the Japanese Registry of Neuroendovascular Therapy 2 (JR-NET2) database and used the information for a retrospective, nation-wide multicenter, observational study to clarify the clinical characteristics, current status of procedures, and outcome of patients treated by neuroendovascular therapy in Japan. In this report, we analyzed the clinical characteristics of dural arteriovenous fistulas (dAVFs) in the JR-NET2 database. All patients with dAVFs treated with endovascular therapy in 150 Japanese hospitals were included. Patient characteristics, clinical presentations, and imaging characteristics were analyzed. A total of 1,075 patients with dAVFs underwent 1,520 endovascular procedures. Of 1,075 patients, 45% were men and 55% were women. The mean age was 65 ± 13 years. The most frequent location of dAVFs was the cavernous sinus (43.6%), followed by the transverse-sigmoid sinus (TSS) (33.4%). Twelve percent of the patients had intracranial hemorrhage, 9% had venous infarction, and 3% had convulsion. The statistically significant independent risk factors of intracranial hemorrhage were TSS, superior sagittal sinus (SSS), tentorium, anterior cranial fossa, cranio-cervical junction, cortical venous reflux (CVR), and varix. Risk factors of venous infarction were age older than 60 years, male sex, TSS, SSS, and CVR. Risk factors of convulsion were male sex, SSS, and CVR. This is the largest nationwide report, to date, of the clinical characteristics of dAVFs treated by neuroendovascular therapy. CVR was a major risk factor of aggressive symptoms.

Key words: dural arteriovenous fistula, neuroendovascular therapy, embolization, characteristics, cortical venous reflux

Introduction

Intracranial dural arteriovenous fistulas (dAVFs) are rare. The crude detection rates of dAVFs were reported to be 0.29 and 0.51 per 1,00,000 adults per year in Japan and Finland, respectively.[1,2] The

frequencies or outcomes of the dAVFs were not reported in the English literature of the nationwide surveys.

DAVFs have been treated with a variety of treatments, including surgeries, endovascular procedures, radiation therapies, and a combination of these treatments. Over the past several decades, there have been rapid technological developments in

microcatheters and embolic materials in the fields of interventional neuroradiology. Combined with experience, these improvements have permitted the application of catheter-based interventions to various types of cerebrovascular disease, and the number of catheter interventions is increasing tremendously in Japan. Currently, most patients with dAVFs are also treated with endovascular procedures.

In this context, a multicenter study group (Japanese Registry of Neuroendovascular Therapy [JR-NET] Study Group) was formed in 2005 to clarify the factors that had an impact on the results of treatment and to establish standardized treatment by catheter intervention and systems of educating operators. This survey consisted of two phases. The first phase was JR-NET, from January 2005 to December 2006, and the second phase was JR-NET2, from January 2007 to December 2009. The target of treatment modalities in JR-NET and JR-NET2 were all neuroendovascular procedures, including embolization of dAVFs.

In 2011, the clinical data of 863 patients with dAVFs from JR-NET data was reported by Kuwayama et al. to clarify the current status of treatment in the Japanese literature.[1] In the present study, we collected a considerable amount of clinical data related to neuroendovascular therapy for cranial dAVFs through the JR-NET2 investigation. We analyzed this data to document the clinical characteristics of dAVFs treated with neuroendovascular therapy in Japan.

Patients and Methods

I. JR-NET2

Patients were derived from the JR-NET2 database. This was a retrospective, multicenter, observational study that took place from January 2007 through December 2009 in 150 neurosurgical centers with 169 neuroendovascular therapy specialists in Japan. A total of 20,854 procedures were included in JR-NET2, which was the largest database of neuroendovascular therapy in Japan. This database included 1,520 procedures (7.29%) with dAVFs.

II. Patient population

We collected all data related to cranial dAVFs from the JR-NET2 database, and analyzed them. All patients were treated with trans-arterial embolization (TAE), trans-venous embolization (TVE), or both. One thousand five hundred and twenty procedures with dAVFs involved embolization. Some dAVFs were treated with multistage procedures, and the patients' background and the clinical manifestation were evaluated in the patients at the time of the

first procedure (patient population). The number of procedures and patient population consisted of 1,520 procedures and 1,075 patients who had undergone or attempted endovascular procedures for dAVFs between January 2007 and December 2009.

III. Evaluation

The dataset of this registry included the following parameters: basic information (facilities, date of treatment, and scheduled or emergent procedures), patient background (age, gender), clinical data [mdified Rankin Scale (mRS) before and 30 days after procedure), complication data (procedure related or not, severity), parameters specific to dAVFs [location, existence of cortical venous reflux (CVR) and varix], and details of endovascular procedures (class of operator, involvement of advising doctor, anesthesia, treatment strategies, type of embolization, type of catheters and embolic materials, result of procedures, and technological success). The locations of dAVFs in this registration were categorized in the nine sites as shown in Table 1.

In this report, we described the epidemiology, such as basic information, patient's background, clinical presentations, and imaging characteristics of each location. Furthermore, we analyzed the risk factors associated with hemorrhage presentation, venous infarction, and convulsion. We plan to give detailed reports separately regarding the details of endovascular procedures and endpoints, such as the mRS score of 0 to 2 at 30 days after treatment, technological success of treatments, and adverse events within 30 days after treatment.

Statistical Analysis

All calculations were performed using JMP 9 software (SAS Institute Inc., Cary, North Carolina, USA). Descriptive statistics were expressed as the means ± standard deviations (SDs). The univariate associations between each potential risk factor and the occurrence of hemorrhage presentation, venous infarction, and convulsion were assessed using Fisher's exact test for comparisons with a cell size less than 10 and Pearson's χ^2 test for others. Results were presented as relative risk (RR) with 95% confidence intervals (CIs). After eliminating variables that were closely related to others, the potential risk factors with a probability value of less than 0.05 on univariate analysis were adopted as confounders in the multivariate logistic regression model for multivariate analysis to determine whether or not risk factors remained independently associated with the occurrence of hemorrhage presentation, venous infarction and convulsion. Results

Table 1 Clinical and imaging characteristics of dural arteriovenous fistulas

	Variable	Total	CS	TSS	SSS	CMS	Tentorium	ACF	SPS	CCJ	Multiple	Others	n.d.
Number of patients		1075	469	359	51	40	31	13	10	10	21	57	14
	Male sex (%)	479 (45)	120 (26)	195 (54)	39 (76)	20 (50)	21 (68)	11 (84)	7 (70)	6 (60)	13 (62)		
	Mean age in yrs ± SD	65 ± 13	67 ± 13	66 ± 11	60 ± 17	62 ± 10	59 ± 14	65 ± 6	63 ± 12	59 ± 16	62 ± 16		
Symptoms	Aggressive symptoms (%)	251 (23)	15 (3)	151 (42)	29 (57)	4 (10)	13 (42)	4 (31)	3 (30)	5 (50)	8 (38)		
	Hemorrhage (%)	129 (12)	6 (1)	74 (21)	12 (24)	2 (5)	10 (32)	4 (31)	2 (20)	4 (40)	3 (14)		
	Venous infarction (%)	93 (9)	7 (1)	63 (18)	10 (20)	1 (3)	3 (10)	0 (0)	1 (10)	0 (0)	4 (19)		
	Convulsion (%)	29 (3)	2 (0.4)	14 (4)	7 (14)	1 (3)	0 (0)	0 (0)	0 (0)	1 (10)	1 (5)		
	Non-aggressive symptoms (%)	677 (63)	446 (95)	145 (40)	12 (24)	33 (83)	5 (16)	2 (15)	4 (40)	3 (30)	10 (48)		
	Asymptomatic (%)	67 (6)	4 (1)	28 (8)	7 (14)	0 (0)	7 (23)	7 (54)	2 (20)	0 (0)	0 (0)		
CVR	CVR w/varix (%)	168 (16)	38 (8)	71 (20)	10 (20)	3 (8)	15 (48)	7 (54)	4 (40)	2 (20)	3 (14)		
	CVR w/o varix (%)	560 (52)	218 (46)	224 (62)	37 (73)	15 (38)	11 (35)	5 (38)	6 (60)	4 (40)	13 (62)		

ACF: anterior cranial fossa, CCJ: cranio-cervical junction, CMS: condylar-marginal sinus, CS: cavernous sinus, CVR: cortical venous reflux, n.d.: not described, SPS: superior petrosal sinus, SSS: superior sagittal sinus, TSS: transverse-sigmoid sinus, w: with, w/o: without, yrs: years.

were presented as odds ratio (OR) estimates of RR with 95% CIs. Significance level was set at a p value of less than 0.05.

Results

I. Location of dAVFs and patient background

Of 1,075 patients, 45% were men and 55% were women. The mean age was 65 ± 13 years (Table 1). In terms of the location of dAVFs, the cavernous sinus (CS) was most frequent. The CS was involved in 469 patients (43.6%), transverse-sigmoid sinus (TSS) in 359 (33.4%), superior sagittal sinus (SSS) in 51 (4.7%), condylar-marginal sinus (CMS) in 40 (3.7%), tentorium in 31 (2.9%), anterior cranial fossa (ACF) in 13 (1.2%), superior petrosal sinus (SPS) in 10 (0.9%), cranio-cervical junction (CCJ) in 10 (0.9%), other locations in 57 (5.3%), unspecified locations in 14 (1.3%), and various locations for multiple lesions in 21 (2.0%). The proportion of men was higher than or equal to 50% for all locations except the CS (26%).

II. Clinical presentation and imaging characteristics

The clinical and imaging characteristics of dAVFs are summarized in Table 1. Two hundred and fifty-one patients (23%) presented with aggressive symptoms. One hundred and twenty-nine patients had intracranial hemorrhage, 93 patients had venous infarction, and 29 patients had convulsion. Six hundred and seventy-seven patients (63%) presented with non-aggressive symptoms. In these patients, 447 patients had ophthalmic symptoms (exophthalmos, chemosis, or ophthalmoplegia), 35 patients had headache, and 195 patients had pulsatile tinnitus or bruit. Sixty-seven patients (6%) were found incidentally, and the symptoms of 24 patients were unknown. One hundred and sixty-eight patients (16%) had dAVF with CVR with varix; 560 patients (52%) had dAVF with CVR without varix; and 320 patients (30%) had dAVF without CVR. Aggressive symptoms and CVR were more abundant in the TSS dAVFs (42% and 82%, respectively) than in the CS dAVFs (3% and 54%, respectively).

The results of univariate and multivariate analysis of factors related to intracranial hemorrhage as a primary symptom are summarized in Table 2. The analysis revealed that TSS location (OR, 4.1, 95% CI, 2.5–6.8; p < 0.0001), SSS location (OR, 4.3, 95% CI, 1.6–9.6; p = 0.0004), tentorium location (OR, 5.8, 95% CI, 2.2–14.6; p = 0.0002), ACF location (OR, 4.1, 95% CI, 1.0–14.7; p = 0.0359), CCJ location (OR, 19.8, 95% CI, 3.8–110.2; p = 0.0004), CVR (OR, 17.5, 95% CI, 5.3–108.5; p < 0.0001), and varix (OR, 3.0, 95% CI, 1.9–4.6; p < 0.0001) were significantly associated with hemorrhagic presentation in patients with dAVFs.

The results of univariate and multivariate analysis of factors related to venous infarction as a primary symptom are summarized in Table 3. The analysis revealed that age older than 60 years (OR, 3.0,

Table 2 Hemorrhagic presentation in dural arteriovenous fistulas

Variable	Univariate			Multivariate		
	RR	95% CI	P value	OR	95% CI	P value
Age older than 60 years	0.9	0.6–1.3	0.6694			
Age older than 70 years	0.8	0.6–1.2	0.415			
Male sex	1.6	1.2–2.3	0.0033	1.2	0.8–1.8	0.5044
Cavernous sinus	0.1	0.0–0.1	< 0.0001			
Transverse-sigmoid sinus	2.7	1.9–3.7	< 0.0001	4.1	2.5–6.8	< 0.0001
Superior sagital sinus	2.1	1.2–3.5	0.0096	4.3	1.6–9.6	0.0004
Condylar-marginal sinus	0.4	0.1–1.6	0.2162			
Tentorium	2.9	1.7–4.9	0.0004	5.8	2.2–14.6	0.0002
Anterior cranial fossa	2.6	1.1–5.9	0.064	4.1	1.0–14.7	0.0359
Superior petrosal sinus	1.6	0.5–5.7	0.3525			
Cranio-cervical junction	3.3	1.5–7.2	0.0252	19.8	3.8–110.2	0.0004
Multiple	1.2	0.4–3.4	0.7355			
Cortical venous reflux	27.8	6.9–111.6	< 0.0001	17.5	5.3–108.5	< 0.0001
Varix	3.9	2.8–5.3	< 0.0001	3.0	1.9–4.6	< 0.0001

CI: confidence interval, OR: odds ratio, RR: relative risk.

95% CI, 1.6–6.0; p = 0.0008), male sex (OR, 2.0, 95% CI, 1.2–3.2; p = 0.0059), TSS location (OR, 4.8, 95% CI, 2.8–8.4; p < 0.0001), SSS location (OR, 4.3, 95% CI, 1.7–10.0; p = 0.001), and CVR (OR, 14.9, 95% CI, 4.6–91.5; p = 0.0002) were significantly associated with venous infarction in patients with dAVFs.

The results of univariate and multivariate analysis of factors related to convulsion as a primary symptom are summarized in Table 4. The analysis revealed that male sex (OR, 3.1, 95% CI, 1.4–8.1; p = 0.0113), SSS location (OR, 4.0, 95% CI, 1.5–9.8; p = 0.0037), and CVR (OR, 9.9, 95% CI, 2.1–178.2; p = 0.0252) were significantly associated with convulsion in patients

Table 3 Venous infarction in dural arteriovenous fistulas

Variable	Univariate			Multivariate		
	RR	95% CI	P value	OR	95% CI	P value
Age older than 60 years	2.4	1.3–4.4	0.002	3.0	1.6–6.0	0.0008
Age older than 70 years	1.7	1.2–2.5	0.0055			
Male sex	2.2	1.4–3.2	0.0001	2.0	1.2–3.2	0.0059
Cavernous sinus	0.1	0.04–0.2	< 0.0001			
Transverse-sigmoid sinus	4.2	2.7–6.3	< 0.0001	4.8	2.8–8.4	< 0.0001
Superior sagital sinus	2.4	1.3–4.4	0.0044	4.3	1.7–10.0	0.001
Condylar-marginal sinus	0.3	0.04–2.0	0.2467			
Tentorium	1.1	0.4–3.4	0.7433			
Anterior cranial fossa	0		0.6196			
Superior petrosal sinus	1.1	0.2–7.3	0.6057			
Cranio-cervical junction	0		1			
Multiple	2.2	1.0–5.4	0.1071			
Cortical venous reflux	20.2	5.0–81.6	< 0.0001	14.9	4.6–91.5	0.0002
Varix	1.4	0.9–2.3	0.1297			

CI: confidence interval, OR: odds ratio, RR: relative risk.

Table 4 Convulsion in dural arteriovenous fistulas

Variable	Univariate			Multivariate		
	RR	95% CI	P value	OR	95% CI	P value
Age older than 60 years	0.7	0.3–1.4	0.2943			
Age older than 70 years	1.0	0.5–2.1	1			
Male sex	3.9	1.7–9.1	0.0009	3.1	1.4–8.1	0.0113
Cavernous sinus	0.1	0.0–0.4	< 0.0001			
Transverse-sigmoid sinus	1.9	0.9–3.8	0.0857			
Superior sagital sinus	6.4	2.9–14.2	< 0.0001	4.0	1.5–9.8	0.0037
Condylar-marginal sinus	0.9	0.1–6.6	1			
Tentorium	0		1			
Anterior cranial fossa	0		1			
Superior petrosal sinus	0		1			
Cranio-cervical junction	3.7	0.6–24.7	0.245			
Multiple	1.8	0.2–12.3	0.4475			
Cortical venous reflux	12.4	1.7–91.0	0.0004	9.9	2.1–178.2	0.0252
Varix	2.0	0.9–4.4	0.1168			

CI: confidence interval, OR: odds ratio, RR: relative risk.

with dAVFs.

Discussion

Two nationwide surveillances of dAVF or vascular malformation were previously published. The report from Scotland was mainly related to arteriovenous malformations and they included only 13 cases with dAVFs.[3] Kuwayama et al.[1] reported the characteristics and status of the treatment of each dAVF in Japan. They included 863 cases with dAVFs treated by endovascular procedures as well as by surgery, radiosurgery, and conservative management. There have been various single and multi-center reported series of dAVFs.[2,4–11] Singh et al.[9] reported the largest series, which included 402 patients with dAVFs. Here, we report the largest survey to date on the characteristics of each dAVF.

I. Location

Kuwayama et al.[1] reported that CS was involved in 45.9% of patients, TSS in 26.7%, spinal cord in 5.9%, anterior condylar confluence in 5.0%, tentorium in 4.8%, SSS in 3.2%, CCJ in 2.4%, cranial vault in 2.4%, anterior cranial base in 2.1%, confluence of the sinus in 1.4%, and multiple locations in 1.4%. On the other hand, the previous largest series from a single center in California reported that CS was involved in 40.5% of patients, TSS in 30.8%, posterior fossa in 8.0%, superior petrosal sinus in 5.5%, SSS in 4.5%, marginal sinus in 4.2%, ethmoidal in 3.0%, middle cranial fossa in 2.2%, inferior petrosal sinus in 1.2%, and multiple locations in 15.9%.

Previous series from Western countries[2,4–6,8,12] reported that the most frequent location of dAVF was TSS, followed by CS. On the other hand, CS was reported as the most frequent location of dAVFs in Asian populations.[1,13,14] Our data confirmed the tendency that CS location is more frequent than TSS location in Asia.

Certain anatomic locations of dAVFs, such as the tentorium, ACF, and CCJ, are more amenable for surgery.[1,15–17] We consider that this explained the low frequencies in tentorium, ACF, and CCJ locations in our survey, which had no dAVF data related to surgery, stereotactic radiation therapy, or conservative cases.

II. Clinical presentation

It is well known that a higher risk of intracranial hemorrhage and non-hemorrhagic neurological deficit is seen in dAVFs with CVR. We reviewed 12 reports related to the risk factors of intracranial hemorrhage and non-hemorrhagic neurological deficit in patients with dAVFs. The results are shown in Table 5.[4,7,9–12,14,18–22] Recently, Singh et al.[9] reported

Table 5   Review of literatures on risk factors for intracranial hemorrhage or aggressive symptoms in dural arteriovenous fistula

Series	Year	No. of cases	Risk factors
Malik et al.[20]	1984	10	Leptomeningeal venous drainage, large variceal dilatation
Viñuela et al.[21]	1986	14	Leptomeningeal venous drainage
Awad et al.[12]	1990	17	Leptomeningeal venous drainage, venous dilatation, galenic drainage
Brown et al.[4]	1994	54	Venous varix
Davies et al.[7]	1996	102	Leptomeningeal venous drainage, sinus occlusion, venous ectasia
Willinsky et al.[22]	1999	130	Pseudophlebitic pattern of venous drainage
Kim et al.[14]	2002	53	Retrograde intracranial venous drainage
van Dijk et al.[11]	2002	236	Persistent cortical venous reflux
Lucas et al.[19]	2006	93	Anterior fossa and tentorial location, leptomenigeal drainage, venous dilatation
Singh et al.[9]	2008	402	Male sex, age, posterior fossa location, cortical venous reflux
Söderman et al.[10]	2008	85	Cortical venous reflux, prsentation with past hemorrhage
Bulters et al.[18]	2012	75	Cortical venous reflux, venous ectasia

the largest series of dAVFs, in which cortical venous drainage, focal neurological deficits, posterior fossa location, male sex, and patients older than 50 years were found to be independently associated with hemorrhagic presentation; CVR has the highest odds ratio (OR, 10.5, 95% CI, 4.9–22.6; p < 0.001). Several major classifications of dAVFs have been developed to grade the risks of dAVFs, including those devised by Cognard et al.,[6] Borden et al.,[23] and Lalwani et al.[8] In our reports, we confirmed the earlier findings that CVR is associated with the risk of intracranial hemorrhage and venous infarction, respectively. Furthermore, we revealed the fact that CVR is associated with the risk of convulsion, too.

The tendency for dAVFs in some locations, such as the tentorium, ACF, SSS, and posterior fossa, to present more frequently with hemorrhage or aggressive symptoms, have been reported.[6,7,9,12] In a previous Japanese survey, Kuwayama et al. reported that the incidence of aggressive symptoms (hemorrhage, venous infarction, elevated intracranial pressure, and convulsion) was 5.6% in CS, 60% in TSS, 67% in confluence, 82% in SSS, 17% in the anterior cranial base, 51% in tentorium, 19% in ACC, 86% in CCJ, and 52% in vault.

We found that intracranial hemorrhage was more abundant in the TSS, SSS, tentorium, ACF, and CCJ locations. Except for the TSS location, our results were similar to those of previous reports. Additionally, we found that venous infarction was abundant in the TSS and SSS locations, and that convulsion was abundant in the SSS location. The dangerous symptoms of these locations are a function of their more dangerous venous anatomies. When major sinuses such as the TSS and SSS are involved in dAVFs with CVR, the influences of blocked venous outflow were greater than that for other sinuses or veins. We believe that this is why patients with dAVFs in the TSS or SSS locations developed more dangerous presentations. The reasons for the low risk of aggressive symptoms in the CS location are the low possession rate of CVR and the high rate of benign symptoms as heralds.

We found that male sex was an independent risk factor for venous infarction and convulsion of dAVFs. In the present study, we demonstrated that CVR was more frequent in men than in women (72% and 64%, respectively), and that the proportion of men was higher than or equal to 50% for all locations except the CS. Previous studies also reported that men had dAVFs with CVR more often than women.[18,24,25] We consider that this prevalence of CVR resulted in a higher risk of venous infarction and convulsion in male patients than in female patients.

III. Study limitations

The present study has some limitations. First, although the amount of data was large, there was no dAVF data related to surgery, radiosurgery, and conservative cases. Thus, our data did not accurately reflect the prevalence of dAVFs. Second, we could not determine if the data was from the same patient or if multiple procedures were done, so we considered the number of first procedures to be the number of patients.

In the future, we need to accumulate all the data of each patient with dAVFs treated by endovascular procedures as well as by surgery, radiosurgery, and conservative management in Japan in order to conduct the definitive nation-wide study of epidemiology.

Conclusion

A total of 1,075 patients with dAVFs underwent 1,520 endovascular procedures between January 2007 and December 2009 in Japan. In terms of location of dAVFs, the CS was the most frequent, followed by TSS. CVR was the major risk factor of aggressive symptoms such as hemorrhagic presentation, venous infarction, and convulsion.

Acknowledgments

Conflicts of Interest Disclosure

The authors declare that they have no conflicts of interest.

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