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Prevalence, Clinical Characteristics, and Hemodynamic Impact of Coronary Artery–Ventricular Microfistulas in Patients with Fontan Circulation

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Background: Coronary artery–ventricular microfistulas (CAMFs), which are defined as diffuse communications between dilated coronary arteries and the main ventricle, are poorly understood in Fontan circulation. We evaluated their prevalence, clinical features, and longitudinal hemodynamics. **Methods:** We retrospectively reviewed 167 Fontan patients undergoing cardiac catheterization between 2005 and 2025. Excluded were sinusoidal communications in PA/IVS, LV fistulas in HLHS, or solitary focal fistulas. Clinical, surgical, and serial hemodynamic parameters, such as the subendocardial viability ratio (SEVR) and aortic regurgitation (AR) index, were assessed. **Results:** CAMFs were identified in 6 of the 153 eligible Fontan patients (prevalence: 3.9%). Polysplenia was more frequent in the CAMFs group (33% vs. 5%, $p = 0.03$). At initial diagnosis, systemic cardiac index (median 3.3 L/min/m² [range, 2.2–3.9 L/min/m²]) and ejection fraction (median 52% [range, 44%–65%]) were preserved. Notably, resting SEVR (median 91% [range, 85%–156%]) and AR index (median 54% [range, 44%–59%]) remained

stable and well-compensated over a decade of follow-up. Late morbidities included protein-losing enteropathy (17%), pulmonary arteriovenous fistulas (17%), esophageal varices (17%), and death (17%). **Conclusions:** CAMFs occur in 3.9% of Fontan patients and correlate with polysplenia. Although resting SEVR and AR index remain well-compensated over mid-term follow-up, their impact on long-term outcomes beyond two decades post-Fontan remains uncertain. Careful long-term monitoring is necessary for late extra-cardiac complications and hemodynamic changes.

Key words: Fontan circulation; Coronary artery anomaly; Microfistula; Subendocardial viability ratio; Polysplenia; Heterotaxy syndrome

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Introduction

The long-term survival of patients undergoing Fontan palliation has improved markedly over recent decades; however, late Fontan circulatory failure and extra-cardiac organ morbidity remain formidable challenges [1]. While coronary artery abnormalities such as myocardial sinusoids in pulmonary atresia with intact ventricular septum (PA/IVS) or ventriculocoronary connections in hypoplastic left heart syndrome (HLHS) are well recognized [2–4], diffuse coronary artery–ventricular microfistulas (CAMFs) are rarely described in single-ventricle lesions [5,6].

CAMFs are characterized angiographically by multiple, diffuse communications arising from dilated, tortuous epicardial coronary arteries and opacifying the main ventricular cavity. Because previous reports are largely limited to isolated case reports, the true prevalence, anatomical associations, longitudinal physiological impact, and long-term prognosis of CAMFs in Fontan patients remain unclarified. Furthermore, whether CAMFs induce diastolic coronary steal and compromise myocardial perfusion balance over time in the Fontan circulation has not been quantitatively evaluated using physiological supply-demand indices.

Therefore, this study aimed to determine the prevalence, clinical characteristics, and long-term hemodynamic impact of CAMFs in a 20-year consecutive cohort of Fontan patients, specifically evaluating myocardial oxygen supply-demand balance using the subendocardial viability ratio (SEVR, also known as the Buckberg index) [7–9] and systemic diastolic pressure preservation via the aortic regurgitation (AR) index [10] through serial mid-term evaluations spanning over a decade post-Fontan.

Methods

Patient Population and Study Design

We conducted a single-center retrospective observational study of 167 consecutive patients with

Fontan circulation who underwent diagnostic cardiac catheterization at Chiba Children's Hospital between January 2005 and August 2025. To specifically investigate diffuse CAMFs, 14 patients were excluded: 11 with sinusoidal communications associated with PA/IVS, 2 with left ventricular microfistulas associated with HLHS, and 1 with a solitary, focal single-vessel coronary artery fistula. The remaining 153 patients were analyzed and categorized into two groups: those with CAMFs (CAMFs group, $n = 6$) and those without CAMFs (non-CAMFs group, $n = 147$). Furthermore, in long-term survivors within the CAMFs group, serial hemodynamic catheterization data (spanning up to 15.0 years post-TCPC) were collected to evaluate longitudinal trajectories of myocardial perfusion and systemic diastolic pressure preservation. This study was approved by the Institutional Review Board of Chiba Children's Hospital with a waived requirement for informed consent via an opt-out protocol.

Angiographic Definition of CAMFs

CAMFs were defined angiographically based on selective coronary angiography or aortic root angiography according to the following specific criteria:

1. Marked dilatation (with tortuosity) and caliber irregularity involving major epicardial coronary arteries.
2. Multiple microfistulous streams arising from each coronary branch draining directly into the main ventricular cavity.
3. Diffuse and widespread opacification of the systemic ventricular lumen during the diastolic phase.

Data Collection and Hemodynamic Evaluation

Demographic parameters, underlying cardiac anatomy, associated syndromes/heterotaxy, surgical history, catheterization hemodynamic parameters, and late Fontan morbidities/mortality were extracted from medical records. Hemodynamic variables recorded included heart rate (HR),

central venous pressure (CVP), systemic ventricular end-diastolic pressure (SVEDP), ascending aortic systolic and diastolic pressures (Sys/Dia AoP), systemic cardiac index (Q_s) calculated via the Fick principle, systemic ventricular ejection fraction (SVEF), pulmonary vascular resistance index (PVR), systemic vascular resistance index (SVR), SEVR, and AR index.

To quantitatively evaluate myocardial oxygen supply–demand balance and systemic diastolic pressure preservation, two specific hemodynamic indices were analyzed:

1. **SEVR:** Calculated from invasive simultaneous aortic and ventricular pressure tracings recorded during cardiac catheterization. SEVR was defined as the ratio of the diastolic pressure-time integral (DTI, reflecting subendocardial coronary perfusion supply) to the systolic pressure-time integral (STI, reflecting myocardial oxygen demand) per cardiac cycle, expressed as a percentage: $\mathrm{SEVR} = (\mathrm{DTI}/\mathrm{STI}) \times 100$. A value of $> 80\%$ was defined as the physiological normal reference threshold [8].
2. **AR Index:** Calculated to assess systemic diastolic pressure runoff and aortic root competence. The AR index was defined as the difference between Dia. AoP and SVEDP divided by Sys. AoP, expressed as a percentage: $\mathrm{AR\ index} = [(\mathrm{Dia.\ AoP} - \mathrm{SVEDP})/\mathrm{Sys.\ AoP}] \times 100$. An AR index $> 25\%$ was considered normal [10].

Statistical Analysis

Continuous variables are expressed as median (interquartile range [IQR]) or mean $\pm$ standard deviation. Categorical variables are reported as frequencies and percentages. Comparisons between the CAMFs and non-CAMFs groups were performed using the Mann–Whitney U test for continuous variables and Fisher's exact test for categorical variables. Longitudinal changes in serial catheterization parameters were evaluated descriptively given the cohort size. A two-tailed p-value of < 0.05 was considered statistically significant. Statistical analyses were performed using JMP 16.0 (SAS Institute Inc., Cary, NC, USA).

Results

Demographic Characteristics and Prevalence

CAMFs were identified in 6 of the 153 eligible Fontan patients, corresponding to a prevalence of 3.9%. Clinical characteristics are summarized in Table 1. There were no significant differences between the CAMFs and non-CAMFs groups regarding age at Fontan completion (median 3.2 vs. 2.5 years, $p = 0.59$), female sex (33% vs. 35%, $p = 0.30$), dominant right ventricular morphology (50% vs. 58%, $p = 0.51$), or presence of underlying genetic syndromes (17% vs. 1%, $p = 0.11$). Heterotaxy syndrome was present in 50% of the CAMFs group versus 35% of the non-CAMFs group ($p = 0.24$). Notably, polysplenia was significantly more frequent in patients with CAMFs than in those without (33% vs. 5%, $p = 0.03$).

Surgical History

Surgical procedures prior to and during Fontan completion were comparable between the two groups (Table 1). Rates of Blalock–Taussig shunt (50% vs. 56%, $p = 0.31$), pulmonary artery banding (17% vs. 23%, $p = 0.38$), Norwood or Damus–Kaye–Stansel anastomosis (33% vs. 20%, $p = 0.25$), bidirectional Glenn or hemi-Fontan procedure (83% vs. 78%, $p = 0.39$), and fenestrated Fontan (33% vs. 24%, $p = 0.30$) showed no statistically significant differences. Pacemaker implantation was performed in 33% of the CAMFs group compared with 6% of the non-CAMFs group, showing a trend toward higher frequency in CAMFs patients ($p = 0.06$).

Hemodynamic Characteristics and Longitudinal Trajectories

Individual catheterization parameters for the 6 CAMFs patients, including serial evaluations, are detailed in Table 2. At initial diagnosis, central venous pressure showed a median of 14.0 mmHg (range, 10–18 mmHg) and systemic ventricular end-diastolic pressure showed a median of 7.0 mmHg (range, 4–11 mmHg). Systemic blood pressure was well preserved, with median ascending

aortic systolic and diastolic pressures of 101.5 mmHg (range, 80–115 mmHg) and 60.0 mmHg (range, 40–75 mmHg), respectively. Systemic cardiac index (median, 3.3 L/min/m²; range, 2.2–3.9 L/min/m²) and ejection fraction (median, 52.0%; range, 44%–65%) were well preserved.

Initial PVR showed a median of 1.7 Wood units $\cdot$ m^2 (range, 1.1–4.9 Wood units $\cdot$ m^2), with an elevated value observed in one patient (Pt 6, 4.9 Wood units $\cdot$ m^2), while initial SVR showed a median of 19.0 Wood units $\cdot$ m^2 (range, 10.5–34.9 Wood units $\cdot$ m^2), with 4 of the 6 patients (67%) exhibiting values below 20.0 Wood units $\cdot$ m^2—including a notably low value reflecting hyperdynamic status in Patient 2 (10.5 Wood units $\cdot$ m^2).

At initial evaluation, the median SEVR among CAMFs patients was 91% (range, 85% to 156%), with all patients maintaining values above the normal physiological threshold of 80%, though 4 of the 6 patients (67%) remained below 100%. Similarly, the initial AR index was well maintained across all patients (median, 54%; range, 44% to 59%), remaining well above the normal threshold of $> 25\%$.

Importantly, serial hemodynamic evaluations in long-term survivors (Pt 5 and Pt 6, spanning up to 15.0 years post-TCPC) demonstrated that resting SEVR (range, 87%–134%), and AR index (range, 45%–51%) remained stable and well within normal ranges over a decade of follow-up. This indicates that effective physiological compensation is maintained at rest despite the presence of CAMFs.

Clinical Morbidity and Outcomes

No acute ischemic cardiac events, angina, or coronary aneurysm ruptures occurred during follow-up. However, patients with CAMFs experienced a high burden of late Fontan-associated morbidities (Table 1). In the CAMFs group, 1 patient (17%) developed protein-losing enteropathy, 1 patient (17%) developed pulmonary arteriovenous fistulas, and 1 patient (17%) developed severe esophageal varices. Ejection fraction $< 45\%$ occurred in 1 patient (17%). One death

(17%) occurred in the CAMFs group secondary to massive esophageal variceal hemorrhage, compared to 9 deaths (6%) in the non-CAMFs group ($p = 0.29$).

Discussion

This study provides several new insights into diffuse CAMFs in the Fontan circulation: (1) CAMFs occur in approximately 3.9% of Fontan patients; (2) CAMFs exhibit a significant association with polysplenia/heterotaxy syndrome; (3) serial evaluations in long-term survivors demonstrate that resting myocardial perfusion balance and systemic diastolic pressure preservation remain stable and well-compensated over mid-term follow-up; and (4) despite preserved resting hemodynamic metrics, CAMFs patients experience a substantial burden of late Fontan circulatory failure and extra-cardiac organ morbidity.

Prevalence and Heterotaxy Predisposition

While coronary communications in PA/IVS or HLHS have been studied extensively, true diffuse CAMFs in single-ventricle physiology are exceptionally rare [2–4]. Finn et al. reported an incidence of 0.8% (2/241) in children with cavopulmonary connections [6]. In our single-center cohort, we established a higher prevalence of 3.9%.

A major finding of our study is the strong statistical association between CAMFs and polysplenia (33% vs. 5%, $p = 0.03$). Heterotaxy syndrome, particularly polysplenia, is inherently characterized by abnormal microvascular development, such as pulmonary arteriovenous malformations and extensive systemic-venous collateral networks [11,12]. Our findings suggest that CAMFs may represent an intra-cardiac vascular manifestation of systemic vascular dysgenesis or the abnormal persistence of embryonic intramyocardial microvascular plexuses intrinsic to heterotaxy phenotypes.

Longitudinal Hemodynamic Stability and Perfusion Dynamics

In classical cardiac physiology, SEVR serves as a validated index of subendocardial oxygen supply and demand, with values $> 80\%$ considered the normal physiological threshold [7–9]. Concurrently, the AR index quantitatively reflects aortic diastolic pressure maintenance and diastolic runoff, with $> 25\%$ establishing normal aortic competence [10].

In Fontan circulation, subendocardial perfusion is theoretically vulnerable due to elevated central venous pressure, which impairs coronary venous drainage, as well as elevated ventricular filling pressures. Diffuse CAMFs could theoretically worsen this vulnerability through a diastolic coronary steal effect, where coronary blood flow bypasses the capillaries and drains directly into the lower-pressure ventricular cavity.

However, a paramount finding of our study is that serial catheterization over mid-term follow-up (up to 15.0 years post-Fontan) demonstrated stability of both resting SEVR (range, 87%–134%) and AR index (range, 45%–51%). Although 4 of 6 patients exhibited initial SEVR values between 85% and 93% (below 100%), all maintained values strictly above the normal physiological threshold ($> 80\%$), with no progressive decline observed over time. In agreement with these stable hemodynamic indices, no clinical acute ischemic cardiac events, angina, or coronary aneurysm ruptures were observed throughout the follow-up period. This longitudinal stability provides functional evidence that resting myocardial perfusion supply-demand balance and systemic diastolic pressure maintenance are robustly compensated over time in the Fontan state, despite the presence of diffuse CAMFs.

Clinical Management and Prognostic Implications

Notably, Patient 2—who had underlying polysplenia—exhibited preserved resting intracardiac hemodynamics (SEVR 156%, AR index 44%, EF 65%) yet suffered a fatal esophageal variceal hemorrhage at age 20. Of note, this patient demonstrated a distinctly low SVR (10.5 Wood units · m^2), reflecting a hyperdynamic circulation. Massive microfistulous shunting, combined with low

SVR, may have induced a chronic high-output state, potentially accelerating Fontan-associated liver disease, portal hypertension, and portosystemic collateral formation.

Furthermore, regarding the pathological characteristics and severe spectrum of CAMFs, Yoshizawa et al. reported an autopsied Fontan case of a 16-year-old girl who suffered a fatal brain hemorrhage caused by a dural arteriovenous fistula [13]. Histologically, their case demonstrated undulating vascular channels within the ventricular myocardium—lined by CD31-positive endothelial cells with sparse smooth muscle and elastic fibers—confirming that CAMFs correspond to residual myocardial sinusoids. In addition, Finn et al. described a patient with polysplenia and univentricular circulation who developed extensive CAMFs at age 18 and ultimately died of fatal pulmonary hemorrhage at age 22 [6]. Although our cohort exhibited stable mid-term hemodynamics, such previously reported fatal neurological or pulmonary complications and unique vascular fragility underscore the potential systemic risks of Fontan-associated microfistulas.

Rather than dismissing CAMFs as benign incidental angiographic findings based on preserved resting hemodynamic indices, it may be prudent to recognize them as a potential marker for adverse vascular remodeling and increased risk for late extra-cardiac organ complications.

Therapeutically, diffuse CAMFs present a distinct dilemma as they are non-amenable to surgical ligation or coil embolization. Similar to non-Fontan adults with multivessel coronary-cameral fistulae where conservative management is favored [14], caution is required in Fontan patients. As suggested by Finn et al., administering non-selective vasodilators may alter microvascular resistance and aggravate diastolic shunting, worsening coronary steal and volume overload [6]. Finally, while resting hemodynamics remained stable over 12.8 years, their impact beyond two to three decades post-Fontan remains uncertain and warrants rigorous long-term observation.

Limitations

There are several limitations to this study. First, because this was a retrospective single-center study on a rare condition, the sample size of the CAMFs group was small ($n = 6$). This limits our statistical power to adjust for potential confounding factors through multivariate analysis. Second, our serial hemodynamic tracking was descriptive and limited to long-term survivors in the CAMFs group, without a matched control group of Fontan patients without CAMFs. Therefore, it remains unclear whether these stable trends are unique to CAMFs patients or typical of certain Fontan subgroups. Third, all hemodynamic evaluations were performed only at rest. Because the main concern with CAMFs is potential coronary steal and reduced coronary flow reserve, the lack of stress testing—such as exercise or drug-induced stress catheterization—means we cannot completely rule out hidden ischemia or perfusion problems during exertion. Finally, while we suggest that low SVR and microfistulous shunting may have caused a high-output state and worsened liver disease or collaterals in Patient 2, this link remains a hypothesis without direct organ blood flow measurements. Larger, multi-center prospective studies using advanced imaging are needed to confirm these findings and clarify the long-term impact of CAMFs.

Conclusions

Diffuse CAMFs occur in approximately 3.9% of Fontan patients and exhibit a significant association with polysplenia/heterotaxy syndrome. Serial evaluations demonstrate that resting subendocardial perfusion and aortic diastolic pressure maintenance remain stable and well-compensated over mid-term follow-up. However, their potential impact on long-term outcomes beyond two decades post-Fontan remains uncertain. Preserved resting hemodynamic metrics should not preclude rigorous long-term clinical surveillance for late extra-cardiac complications and progressive Fontan failure.

Table 1. Demographic, Surgical, and Clinical Characteristics of Fontan Patients With and Without Coronary Artery–Ventricular Microfistulas (CAMFs).

Continuous data are presented as median (interquartile range), and categorical variables are presented as number (percentage). Comparisons between the CAMFs and non-CAMFs groups were performed using the Mann–Whitney U test for continuous variables and Fisher's exact test or the chi-square test for categorical variables.

Abbreviations: AV, arteriovenous; CAMFs, coronary artery–ventricular microfistula; DKS, Damus–Kaye–Stansel; EF, ejection fraction.

Table 2. Individual Hemodynamic Profile and Longitudinal Catheterization Data of Patients With Coronary Artery–Ventricular Microfistulas (CAMFs).

AR index was calculated as the difference between ascending aortic diastolic pressure and systemic ventricular end-diastolic pressure divided by ascending aortic systolic pressure, expressed as a percentage. SEVR was calculated as the ratio of the diastolic pressure-time integral (DTI) to the systolic pressure-time integral (STI) per cardiac cycle, expressed as a percentage.

Abbreviations: AR index, aortic regurgitation index; Cathe, catheterization time point; CVP, central venous pressure; Dia. AoP, diastolic ascending aortic pressure; HR, heart rate; Pt, patient; PVR, pulmonary vascular resistance index; Q_S, systemic cardiac index; SEVR, subendocardial viability ratio; SVEDP, systemic ventricular end-diastolic pressure; SVEF, systemic ventricular ejection fraction; SVR, systemic vascular resistance index; Sys. AoP, systolic ascending aortic pressure; TCPC, total cavopulmonary connection.

Figure 1. Representative angiographic features of coronary artery–ventricular microfistulas (CAMFs).

(A, B) Aortic root angiography in Patient 1 (A: anteroposterior view, B: lateral view) and (C, D) Patient 2 (C: anteroposterior view, D: lateral view), illustrating diffuse, multiple microfistulous communications arising from the branches of the dilated coronary arteries and draining directly into the main ventricular cavity, resulting in widespread opacification of the systemic ventricular lumen.

Abbreviations: CAMFs, coronary artery–ventricular microfistulas.

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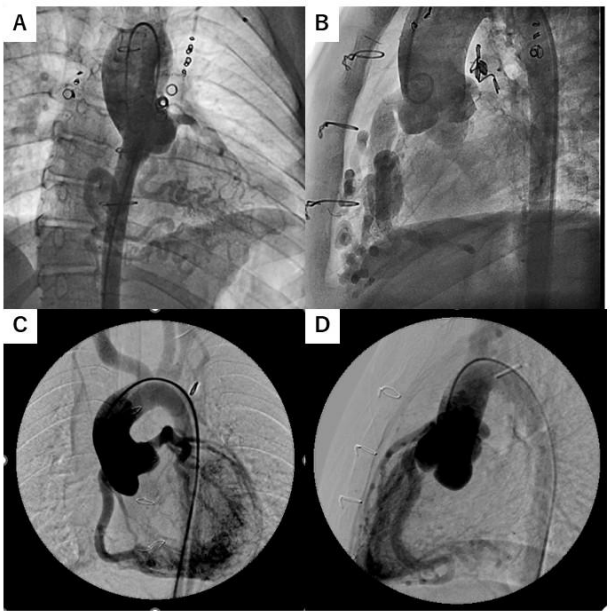


Figure 1

1

	Total (N = 153)	CAMFs group (n = 6)	Non-CAMFs group (n = 147)	p-value
Age at Fontan completion, years	2.5 (1.9–3.2)	3.2 (2.1–3.5)	2.5 (1.9–3.2)	0.59
Female sex	53 (35%)	2 (33%)	51 (35%)	0.30
Right ventricular predominance	88 (58%)	3 (50%)	85 (58%)	0.51
Genetic syndrome	3 (2%)	1 (17%)	2 (1%)	0.11
Heterotaxy syndrome	54 (35%)	3 (50%)	51 (35%)	0.24
Asplenia	45 (29%)	1 (17%)	44 (30%)	0.31
Polysplenia	9 (6%)	2 (33%)	7 (5%)	0.03
Surgical Procedures				
Blalock–Taussig shunt	85 (56%)	3 (50%)	82 (56%)	0.31
Pulmonary artery banding	35 (23%)	1 (17%)	34 (23%)	0.38
Aortic arch repair	9 (6%)	0 (0%)	9 (6%)	0.69
Norwood or DKS anastomosis	31 (20%)	2 (33%)	29 (20%)	0.25
Bidirectional Glenn / Hemi-Fontan	120 (78%)	5 (83%)	115 (78%)	0.39
Fenestrated Fontan	37 (24%)	2 (33%)	35 (24%)	0.30
Pacemaker implantation	11 (7%)	2 (33%)	9 (6%)	0.06
Morbidity and Mortality				
Ruptured coronary aneurysm	0 (0%)	0 (0%)	0 (0%)	1.00
Systemic ventricular EF < 45%	8 (5%)	1 (17%)	7 (5%)	0.25
Protein-losing enteropathy	17 (11%)	1 (17%)	16 (11%)	0.38
Pulmonary AV fistula	3 (2%)	1 (17%)	2 (1%)	0.11
Esophageal varices	3 (2%)	1 (17%)	2 (1%)	0.11
All-cause mortality	10 (6%)	1 (17%)	9 (6%)	0.29

Table 1

Patient	Cathe	Age (yrs)	TCPC Duration (yrs)	HR (bpm)	CVP (mmHg)	SVEDP (mmHg)	Sys/Dia AoP (mmHg)	Qs (L/min/m²)	SVEF (%)	PVR (Wood units-m²)	SVR (Wood units-m²)	AR index (%)	SEVR (%)	Morbidity
Pt 1	Initial Diagnosis	3.9	0.6	110	18	9	95 / 65	3.9	52	1.3	18.3	59	86	Alive
Pt 2	Initial Diagnosis	9.9	6.4	60	11	5	80 / 40	3.9	65	1.1	10.5	44	156	Death at age 20 yrs (Variceal hemorrhage)
Pt 3	Initial Diagnosis	10.5	8.7	90	13	7	115 / 74	3.3	44	1.8	19.7	58	88	Alive
Pt 4	Initial Diagnosis	13.3	10.2	95	15	10	115 / 75	2.5	52	2.0	34.9	57	93	Alive
Pt 5	Initial Diagnosis	6.3	0.8	85	10	4	91 / 50	3.2	54	1.5	16.9	51	105	Alive
	Follow-up 1	15.7	10.2	70	14	8	84 / 51	2.5	51	3.0	22.8	51	134	
Pt 6	Initial Diagnosis	3.9	2.2	105	16	6	108 / 55	2.2	51	4.9	29.5	45	87	Alive
	Follow-up 1	12.3	10.6	80	14	9	112 /65	3.0	54	1.5	26.1	50	88	
	Follow-up 2	16.7	15.0	80	10	8	116 / 66	2.9	54	2.7	28.3	50	112	

Table 2

1