Usefulness of Balloon Angioplasty for the Right Ventricle-Pulmonary Artery Shunt with the Modified Norwood Procedure

Naoki Ohno, MD[1]
Shinichi Ohtsuki, MD, PhD[1]
Koichi Kataoka, MD, PhD[1]
Kenji Baba, MD, PhD[1]
Yoshio Okamoto, MD[1]
Maiko Kondo, MD[1]
Shunji Sano, MD, PhD[2]
Shingo Kasahara, MD, PhD[2]
Osami Honjo, MD, PhD[2]
Tsuneo Morishima, MD, PhD[1]

Department of Pediatrics[1], Department of Cardiovascular Surgery[2]
Okayama University Graduate School of Medicine, Dentistry and Pharmaceutical Sciences, Okayama, Japan

Correspondence for review and proofreading
Dr. Naoki Ohno, MD
Division of Cardiology, The Hospital for Sick Children
555 University Avenue, Toronto, M5GX8,
ON, Canada
Email:naoki.ono@sickkids.ca

Correspondence for reprint
Ohtsuki S, MD, PhD, Okayama University Graduate School of Medicine, Dentistry and Pharmaceutical Sciences, Department of Pediatrics, 2-5-1 Shikata-cho, Kitaku, Okayama, 700-8558, Japan
Email: tsuki33@cc.okayama-u.ac.jp

Keyword: Balloon Angioplasty, RV-PA shunt, Hypoplastic Left Heart Syndrome, Norwood

Short title: Balloon Angioplasty for the RV-PA shunt

Word count:3075

Abstract (Word count: 225)

Objective: We sought to evaluate the efficacy of balloon angioplasty (BA) for severely desaturated patients due to a stenotic right ventricle (RV) to pulmonary artery (PA) shunt following modified Norwood procedure.

Methods: Of 87 patients who underwent a Norwood procedure with the RV-PA shunt between February 1998 through March 2010, 22 (25%) patients underwent BA. The efficacy of BA was assessed by angiographic measurement of the changes in the internal diameters of the stenotic portions of the shunt, changes in arterial saturation and clinical outcomes.

Results: BA was performed for stenotic RV-PA shunts following stage I palliation (n=17, 77%), or those placed as an additional blood source (n=5, 23%, 3 patients awaiting biventricular repair, 2 patients following stage II palliation). The location of the BA was at the distal anastomosis in 12 (54.5%), proximal anastomosis in 21 (95.4%) and in the mid-portion of the shunt in 11 (50%) cases. The diameters of these 3 shunt portions were measured from the anterior-posterior and lateral angiographic images, increasing significantly after BA ($p<0.0001$) in all. Arterial saturation significantly improved after BA in all cases (66.5±4.3% to 79.4±3.4%, $p<0.0001$). Freedom from reintervention was 100%. All patients underwent subsequent elective planned surgery at an appropriate age with no mortality.

Conclusions: A BA-alone strategy for a stenotic RV-PA shunt was effective for all 3 shunt portions, minimizing shunt-related premature surgical intervention.

BACKGROUND

Since Norwood and colleagues discribed neonatal open-heart palliation for patients with hypoplastic left heart syndrome (HLHS)(1), the clinical outcome of this procedure has improved dramatically(2, 3). One recent modification, the use of a right ventricle (RV)-to-pulmonary artery (PA) shunt instead of a modified Blalock-Taussig (BT) shunt in stage I palliation, appears to offer more stable hemodynamics by avoiding diastolic run off(4). A recent randomized trial comparing the two shunt types for stage I palliation showed a significantly higher 1-year transplant-free survival in patients with the RV-PA shunt than those with a BT shunt(5). However, it also revealed a significantly higher requirement for reintervention in the RV-PA shunt group prior to stage II palliation compared to the BT shunt group(5).

Early stenosis of the RV-PA shunt following stage I palliation has been a well-documented phenomenon (6-10). The mechanisms of stenosis include kinking or compression of the shunt, distal anastomotic stenosis with or without branch PA stenosis, proximal stenosis related to muscular growth or endothelial growth within the shunt lumen. As such, progressive desaturation between stages mandates either early conversion to bidirectional Glenn (BDG) anastomosis or catheter intervention to a stenotic RV-PA shunt. Although there are a few case studies describing the efficacy of stent placement in the RV-PA shunt(6,11-14), limited data exists regarding efficacy and durability of balloon angioplasty (BA) alone. In this study, we reviewed our institutional experiences of BA on the RV-PA shunt focusing on the effectiveness of the procedure, reintervention and subsequent timing of stage II palliation.

SUBJECTS AND MATERIALS

Between February 1998 to March 2010, 87 infants underwent the Norwood procedure with a RV-PA shunt at Okayama University Hospital. The subjects of this study were 22 (25.2%) of the 87 patients who had RV-PA conduit stenosis and severe desaturation.

Surgical techniques and institutional policy

The technique of modified Norwood procedure with a RV-PA shunt has been described elsewhere(4,15). Briefly, the RV-PA shunt [expanded polytetrafluoroethylene (ePTFE), Gore-Tex, W.L Gore & Associates, Flangstaff, AZ] was created with a distal cuff to minimize the distal anastomotic stenosis and/or branch PA stenosis. All shunts were anastomosed to the stump of the central branch PA, which was inserted into the left side of the reconstructed aorta. A 5 mm ePTFE was used in the majority (20/22 patients, 90%), and 6 mm shunt was used in 2 patients. In 7 cases with excessive pulmonary blood flow, a hemoclip was placed on the RV-PA shunt to restrict the pulmonary blood flow either intraoperatively or at the time of delayed chest closure. The Norwood procedure was used as initial palliation for potential biventricular repair in 3 patients and at stage II palliation, the RV-PA shunt was left in place providing an additional pulmonary blood flow in 17 partients.

Our institutional policy is to perform stage II palliation at 6 months of age with a body weight of more than 5 kg. Therefore, we used a strategy to treat severely desaturated patients following stage I palliation with a BA. Indications for BA was severe desaturation (arterial saturation less than 70%) clearly related to a RV-PA shunt stenosis. The patients with severe desaturation were investigated by echocardiography [α10 (ALOKA), Vivid 7 (GE Yokogawa Medical Systems), IE33 (PHILIPS)]. Presence of stenosis was evaluated by morphology and flow velocity near and inside the shunt.

Catheterization techniques

All procedures were performed under general anesthesia. Measurement of the PA pressure was often not performed as the patients were unstable. Biplane angiography was performed using either an INTEGRIS BH5000 (PHILIPS) or Infinix Celeve-I

INFX-8000V (TOSHIBA) angiographic system. Several different balloons were used including Wanda, Sasuga, Sterling (Boston Scientific, Natic, MA, USA) or Lacross (GOODMAN CO., LTD, Gifu, Japan) of 5 or 6 mm in diameter and 15 to 40 mm in lengths. A 5mm balloon was used in the 5mm ePTFE shunts, while a 6mm balloon used in the two 6mm shunts. The RV-PA shunt was approached from the femoral vein in all cases. A 4-French Judkins right coronary catheter was used to cannulate the conduit, a 0.014”-0.018” inch guide wire was then placed in one of the PA branches. The balloon was inserted into the shunt as quickly as possible and inflated. Multiple inflations were often performed until the waist of the balloon at the stenotic site disappeared. Subsequent effectiveness of BA was evaluated based on increase in the internal diameter of the shunt on the post-BA angiogram and also an increase in the arterial saturation. Imaging was performed again after BA and the diameter of the stenotic portion was measured. Typical images for each of three portions, before and after BA are shown in **Figure 1**.

Angiographic evaluation

The image analysis software Elk C. View version 1.7 (Elk) was used for measurement of the diameter of the stenotic portions. All original angiographic images were retrospectively analyzed off line. Sites of stenosis were divided into 3 groups: RV-PA shunt connection to the central branch PA (distal portion), site of RV-PA shunt connection to the RV (proximal portion), and inside the RV-PA shunt (inside portion). The angiograms were obtained in the anterior-posterior (AP) and lateral projections, with the children in isocenter for magnification correction. The diameter of the stenotic portion was measured independently and compared before and after BA.

Clinical evaluation

We analyzed data regarding age at the time of BA, body weight, changes in arterial saturation before and after BA, and subsequent postoperative course including time to the next operation.

Statistical Analysis

SPSS Ⅱ (IBM, United States) was used for statistical analyses. A paired T test was used for comparison of data before and after BA and $p<0.05$ was considered statistically significant.

RESULTS

Patient characteristics

Seventeen patients (77%) underwent BA following stage I palliation for single ventricle palliation. The mean age and body weight was 137 days and 4.9 kg, respectively. Five patients had the RV-PA shunt as an additional source of pulmonary blood flow: 3 awaiting biventricular repair in whom a BT shunt (4mm) had already been placed and 2 patients following stage II palliation. The mean age and mean weight of these patients was 547 days and 7.7 kg, respectively.

Efficacy of Balloon Angioplasty

BA was performed at the distal anastomosis in 12 (54.5%) , at the proximal anastomosis in 21 (95.4%) and inside the RV-PA shunt in 11 (50%) cases. Eighteen (82%) patients had multiple stenoses. Of 11 patients who underwent BA for the stenosis inside the shunt, seven had a previous hemoclip placed at stage I palliation. The mean fluoroscopy and total procedure times were 28 minutes and 173 minutes, respectively.

The internal diameters of the RV-PA shunts increased significantly after BA in all 3 portions ($p<0.0001$ for all)(**Figure 2**). In all cases, arterial saturation also significantly improved (66.5±4.3% to 79.4±3.4%, $p<0.0001$). During the procedure, transient arrhythmias occurred in 2 cases with no hemodynamic compromise. There were no other complications during the procedures.

Clinical outcomes

All 17 patients who underwent BA following stage I palliation had a stable arterial saturation (75.6±2.5%) with favorable weight gain. None of the patients required further catheterization or surgical intervention and underwent an elective BDG with no mortality. The mean age and body weight at surgery was 198 days and 5.6 kg, respectively. Six out of 17 patients completed a TCPC and the remaining 11 patients are awaiting a TCPC completion with no late mortality.

Among the 5 patients who had an RV-PA shunt as an additional source of pulmonary blood flow, subsequent biventricular repair was performed in 3 with no mortality with preoperative arterial saturations of 74.3±2.5%. Of the 2 patients who underwent BA following a BDG, one patient underwent TCPC completion (arterial saturation 78% and body weight 11 kg at the age of 4 years) and the other patient is awaiting TCPC completion (arterial saturation 77%). As a whole, none of the patients experienced desaturation episodes after BA. (freedom from reintervention, 100%).

DISCUSSION

Early stenosis of the RV-PA shunt following the modified Norwood procedure is a critical factor contributing to inter-stage mortality and morbidity(3,5). Progressive desaturation during this period necessitates urgent intervention to re-establish adequate pulmonary blood flow in order to avoid a premature second stage procedure. Our results clearly show that BA was effective in dilating all three portions of stenosis without major complications. Arterial saturations increased significantly and remained stable through the next planned surgical intervention, despite having no stent placed with in the shunts. In addition, this strategy worked well for three different clinical situations, i.e., following stage I palliation, an RV-PA shunt as additional pulmonary blood source following stage II palliation and for those having future biventricular repairs.

Balloon Angioplasty for the Distal Anastomotic Stenosis

Ino et al. suggested the main mechanism of balloon dilatation for a stenotic PA was due to non-stretch mechanisms such as tearing, flap formation or dissection(16). The same mechanism may apply if a RV-PA shunt is anastomosed to the native PA without patch augmentation on the central branch PA. In our series, a large ePTFE cuff was pre-anastomosed to the RV-PA shunt. Therefore, there is a large area of ePTFE at the distal anastomosis, which may have made BA more amenable on this particular lesion.

In other words, the balloon may have stretched mainly the ePTFE graft and cuff rather than actual native PAs.

Balloon Angioplasty for Proximal Anastomotic Stenosis

Given the nature of the proximal anastomosis, where the shunt takes off with a sharp angle from the small right ventriculotomy, the proximal anastomosis was a common stenotic site in our series (95%). Potential mechanisms include a kinking of the shunt in the AP direction, muscle growth at the ventriculotomy or fibromuscular proliferation into the shunt. We speculated that BA might not be effective at dilating in the AP direction due to the structural kinking, however, our results showed that BA dilated the shunt in the lateral direction rather than in the AP direction.

Balloon Angioplasty for In-Stent Stenosis

The majority (7/11, 63%) of in-stent stenosis in this series was ‘intentional’, created by a hemoclip placed at the time of stage I palliation to control pulmonary blood flow. Progression of desaturation was expected for those patients and therefore more meticulous follow-up was implemented. We believe that this strategy represents an effective means to manipulate the amount of pulmonary blood flow through the RV-PA shunt, thereby preventing sudden circulatory collapse due to pulmonary overcirculation in the immediate postoperative period. The mechanism of stenosis in the remaining 4 patients was not related to hemoclip placement, and may be due to tissue proliferation associated with the procedure and/or minor kinking of the shunt. BA was also effective for this lesion partly because of the nature of ePTFE since the material itself is expandable by pressure(17).

Balloon Angioplasty vs. Stent Placement

A large number of reports suggest stent placement efficatious for stenotic RV-PA shunts following stage I palliation(6,7,11-14). Re-intervention rates after stent placement are low. Potential disadvantages of stent placement include: distortion of the branch PA bifurcation, erosion of the anastomotic sites, in-stent stenosis and complications related to stent removal at the time of the next operation. None of these potential complications became an issue using a BA-alone strategy. If BA is not effective to improve the internal diameter and/or arterial saturation, stent placement or early surgical intervention can be considered as the future options.

Balloon Angioplasty Following Stage I palliation

Even though a substantial body of evidence shows that early stage II palliation can be safely performed as early as the age of 3 months (18), we still believe that the physiology of cavopulmonary connection can be achieved in the safest manner when the pulmonary vasculature is fully matured. In addition, it is possible that a stenotic RV-PA shunt does not supply sufficient pulmonary blood flow to facilitate normal growth of peripheral pulmonary vasculature, leaving premature lung vasculature with or without a vulnerable pulmonary vascular resistance. Therefore, we think that gaining an additional 2 to 3 months by BA could be beneficial. All 17 patients underwent a BDG at the standard time and had favorable postoperative clinical outcomes.

Balloon Angioplasty for RV-PA shunt as an Additional Pulmonary Blood Source

We have a rather unique surgical strategy, where a RV-PA shunt is left in place at the time of a BDG, serving as an additional pulmonary blood source. This shunt may be beneficial in terms of facilitating further native PA growth and potentially preventing development of arteriovenous fistula by providing hepatic factors. However, we do recognize that the presence of an additional pulmonary blood source following stage II palliation is controversial. Our limited experience (2 patients) showed that BA for RV-PA shunt stenosis following BDG was effective and also in patients who were awaiting biventricular repair(19).

Study Limitation

The major limitations of this study are due to the retrospective non-randomized nature of the patient cohort. Since our institutional policy has been consistent during the study period, there is no comparison group. Lastly, there were no follow-up angiograms to document the medium-term results of BA. Our measure of success comes from immediate post-operative angiographic results and clinical outcomes.

CONCLUSIONS

The BA-alone strategy for stenotic RV-PA shunts in severely desaturated patients was effective in all three shunt portions. Premature surgery was avoided and all patients underwent elective operations at an appropriate age with good clinical outcomes. This strategy can be considered effective to minimize shunt-related premature surgical intervention.

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LEGENDS FOR FIGURES

Figure.1

Figure shows the typical images for each of three portions, before and after BA. A: Distal portion, pre BA, B: Proximal portion pre BA, C: In-stent pre BA, D: Distal portion, post BA, E: Proximal portion, post BA, F: In-stent, post BA.

Figure2

The changes in the internal diameters before and after BAP. A: Comparison of distal portion in the AP image (2.7±0.6 vs. 3.7±0.3mm), B: Comparison of distal portion in the lateral image (3.0±0.6 vs. 4.0±0.4mm), C: Comparison of proximal portion in the AP image (2.9±0.6 vs. 4.0±0.6mm), D Comparison of proximal portion in the lateral image (2.7±0.5 vs. 3.6±0.4mm), E Comparison of inside portion in the AP image (3.1 ±0.8 vs. 4.2±0.7mm), F Comparison of inside portion in the lateral image (2.9±0.7 vs. 4.1±0.4mm)

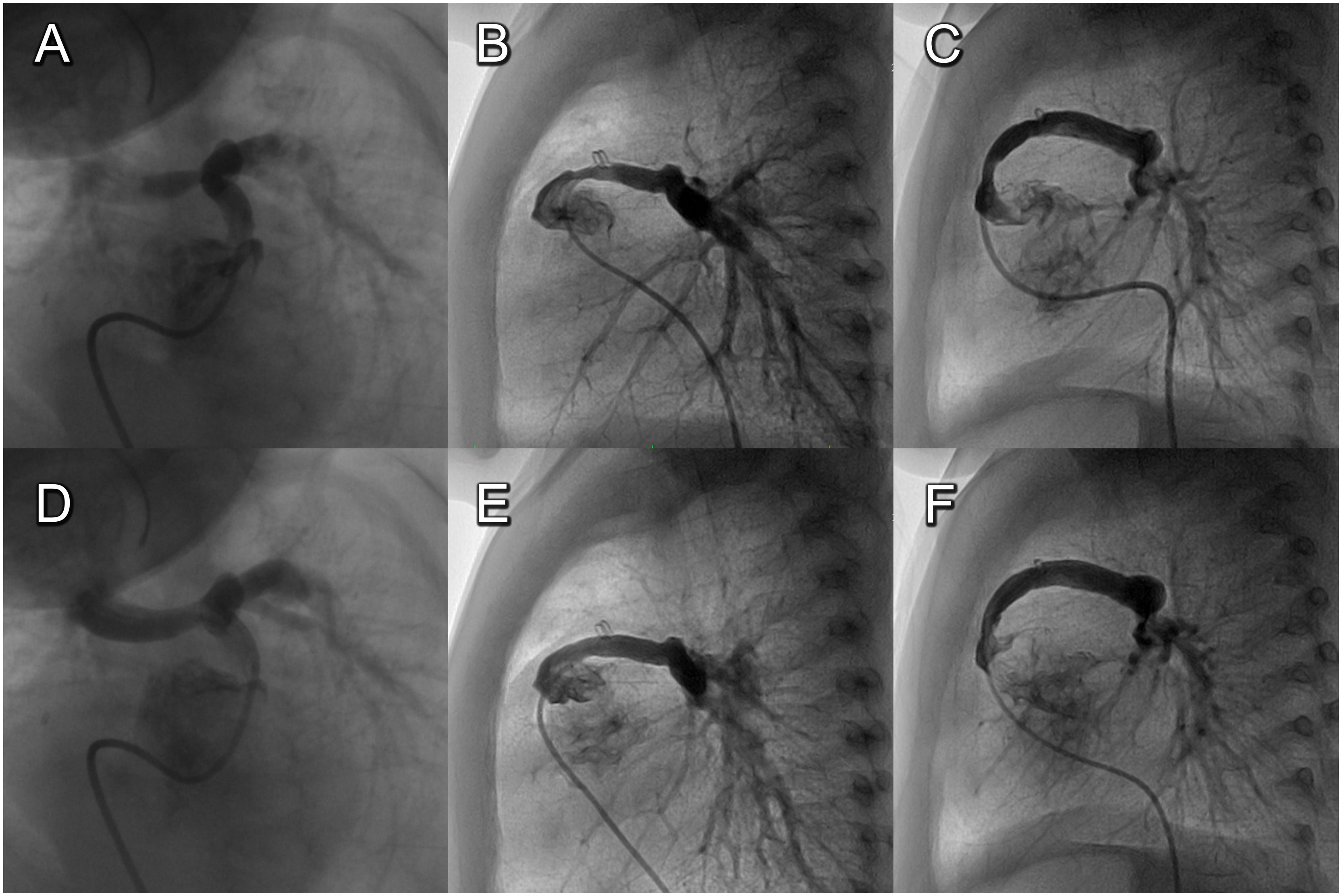

Figure 1.

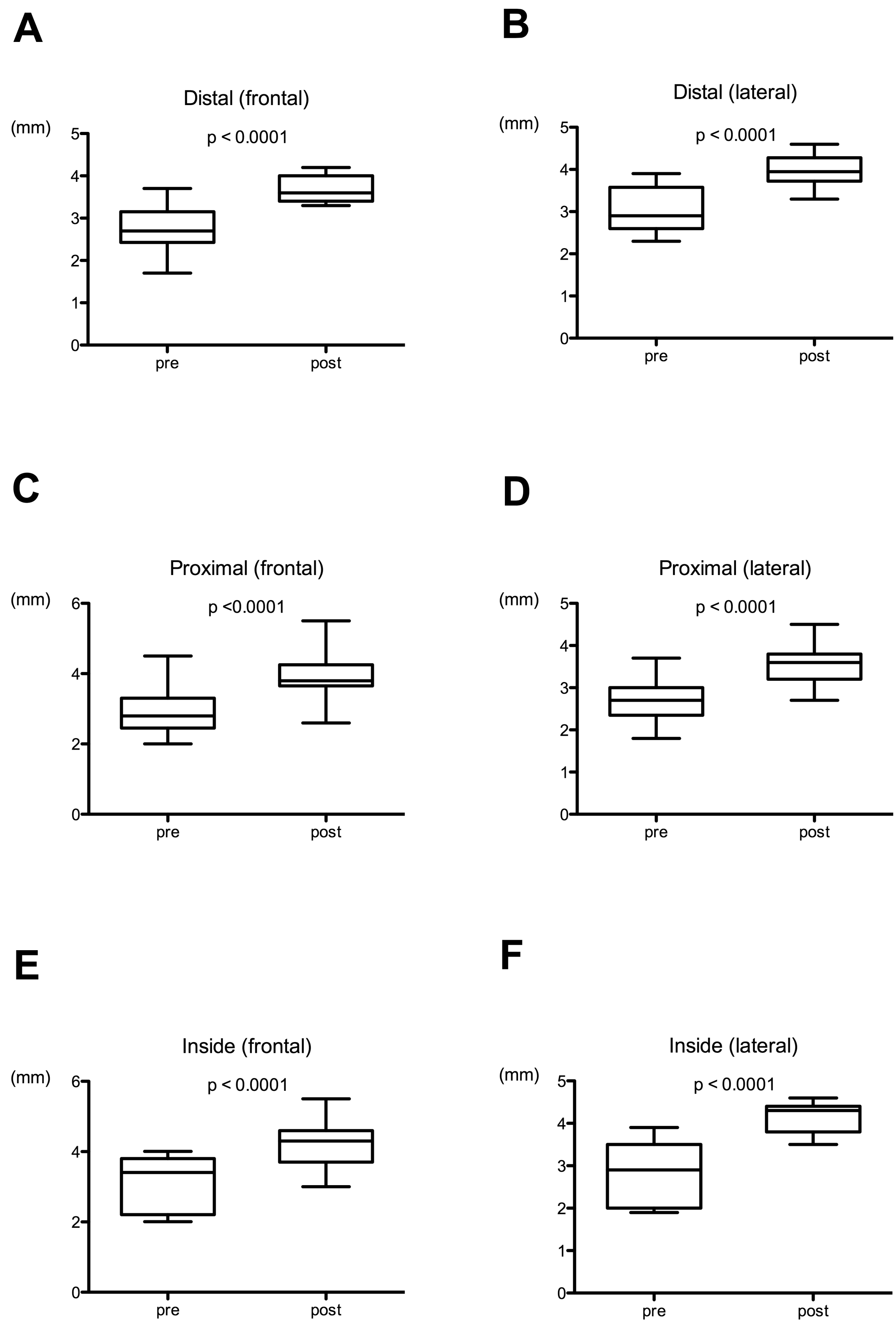
A
Distal (frontal)
(mm)
p < 0.0001
pre
post
B
Distal (lateral)
(mm)
p < 0.0001
pre
post
C
Proximal (frontal)
(mm)
p <0.0001
pre
post
D
Proximal (lateral)
(mm)
p < 0.0001
pre
post
E
Inside (frontal)
(mm)
p < 0.0001
pre
post
F
Inside (lateral)
(mm)
p < 0.0001
pre
post

Figure 2.