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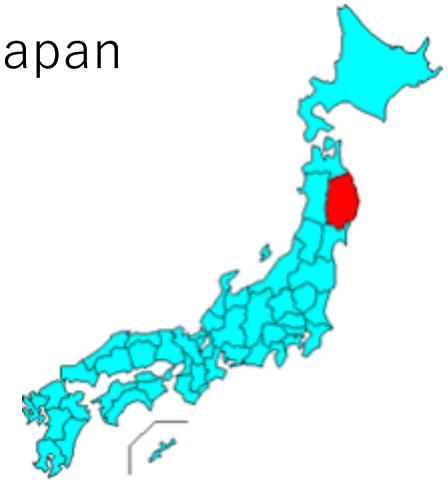
A Seventeen-Year-Old Boy with Left-Dominant Unbalanced AVSD, TOF and Trisomy 21, who Successfully Underwent a Biventricular Conversion: Case Report

Department of Cardiovascular Surgery, Iwate medical University, Japan

Takuya Goto, Junichi Koizumi, Tatsunori Tsuji,

Kazuki Yakuwa, Hajime Kin



AAPCHS

Iwate Medical University 岩手医科大学

Conflict of Interest (COI) of the Principal Presenter
: No potential COI to disclose

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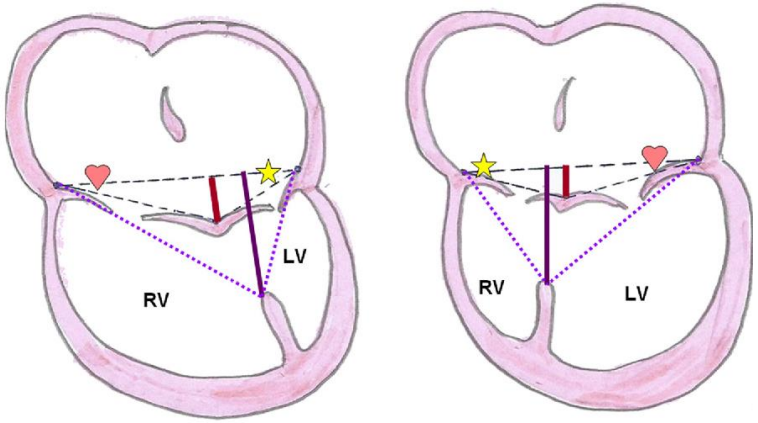
Backgrounds

Unbalanced atrioventricular septal defect (UAVSD)



Right dominant　　Left dominant

The Journal of Thoracic and Cardiovascular Surgery
Volume 153, Issue 2, February 2017, Pages 430-438

Single ventricle management

Challenging to apply single ventricular circulation

Developing pulmonary hypertension

Trisomy 21 (Down syndrome) patients

The Annals of Thoracic Surgery
Volume 101, Issue 5, May 2016, Pages 1834-1841

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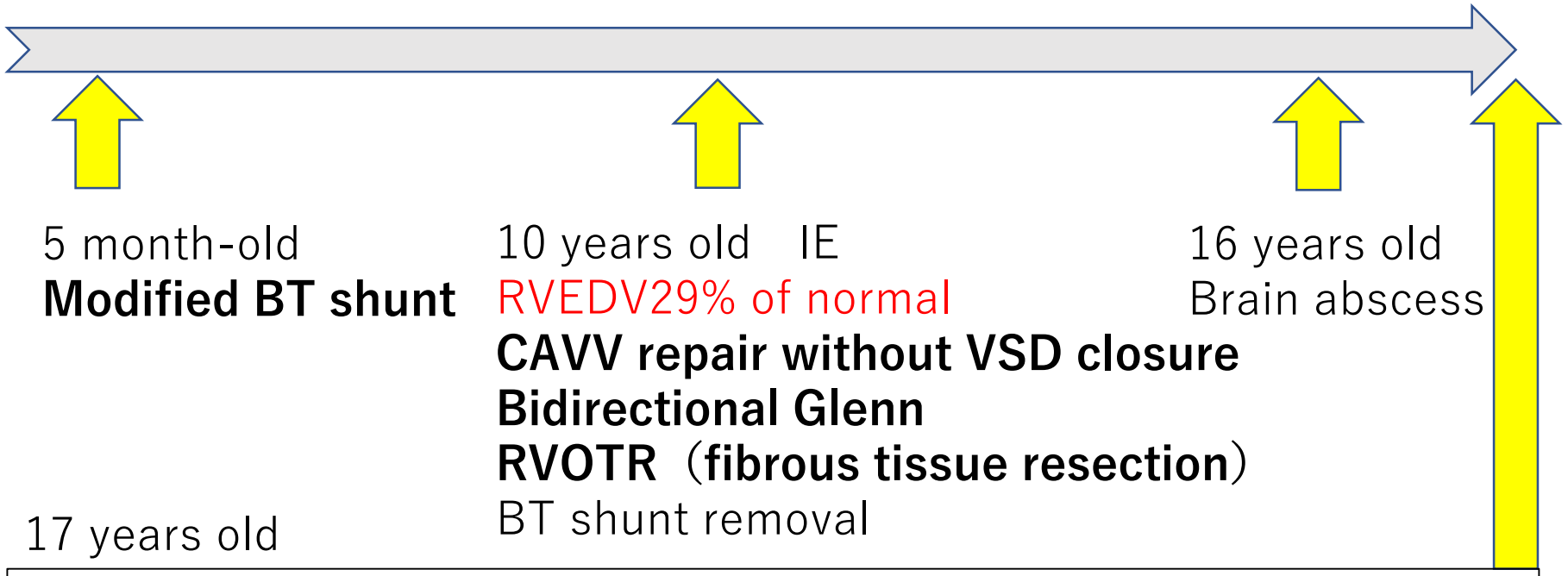
Case

- Male, 17 years old, left dominant unbalanced AVSD, TOF, trisomy 21



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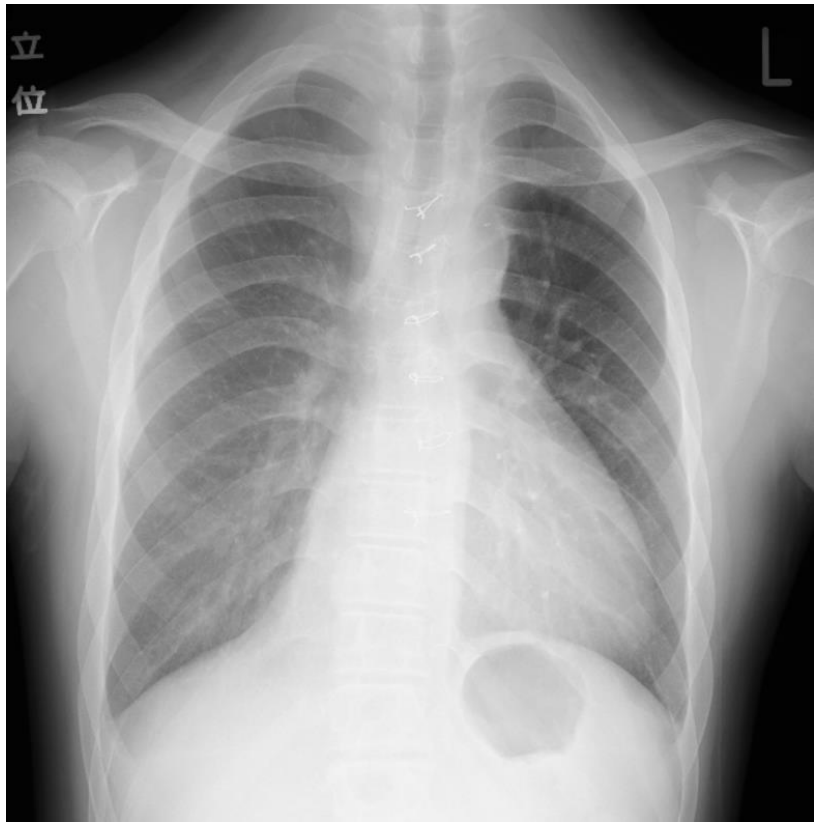
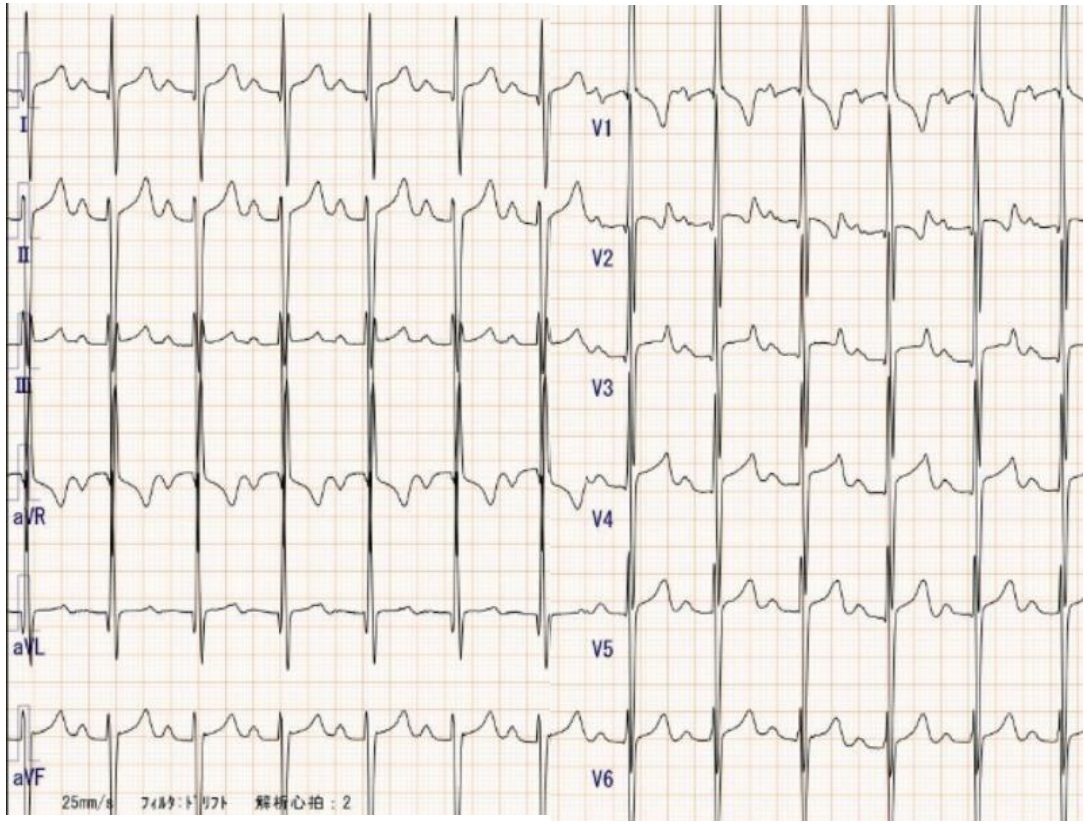
X ray、ECG





- CTR 59%
- CRBBB, RVH

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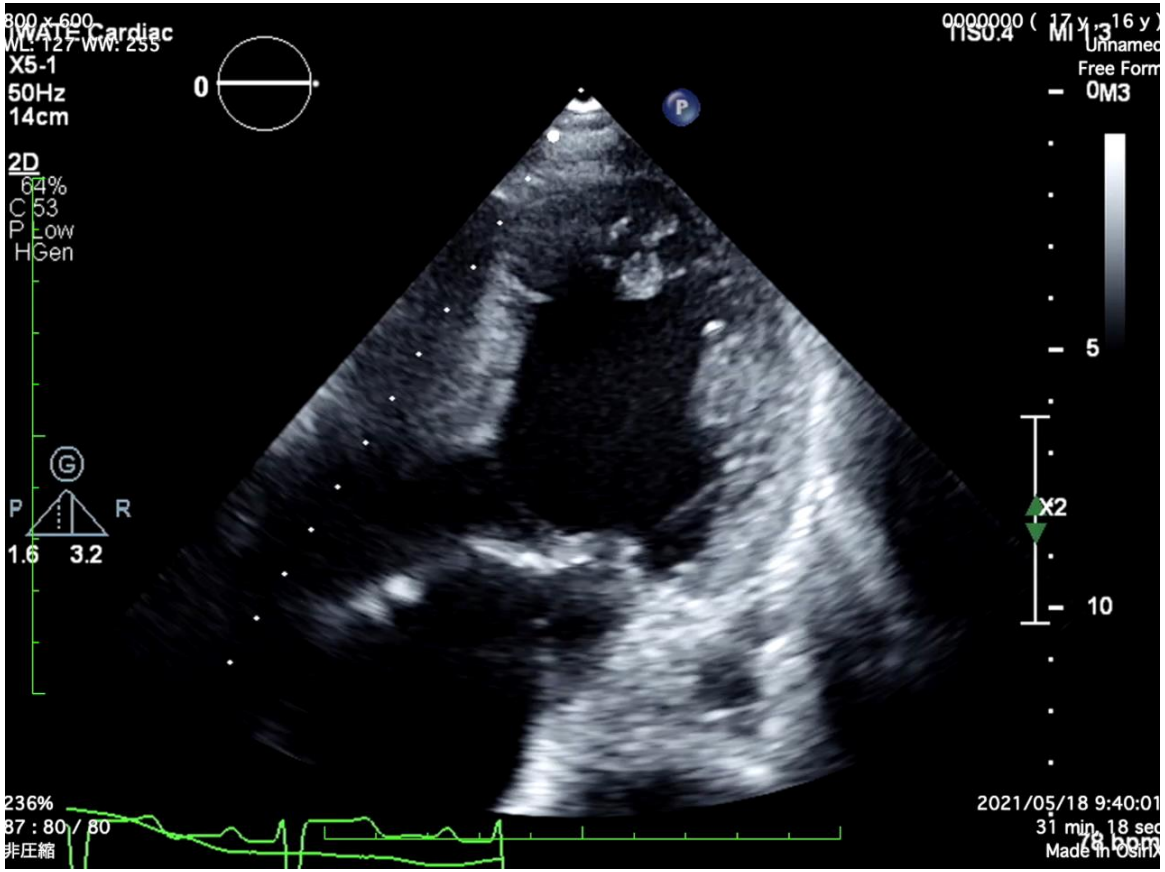
Preoperative echocardiogram



LVEDd 119% of normal
LVEF50%

TAPSE　23㎜
RV FAC　34%

CAVVR moderate
AR mild
RVOT 3.9m/s

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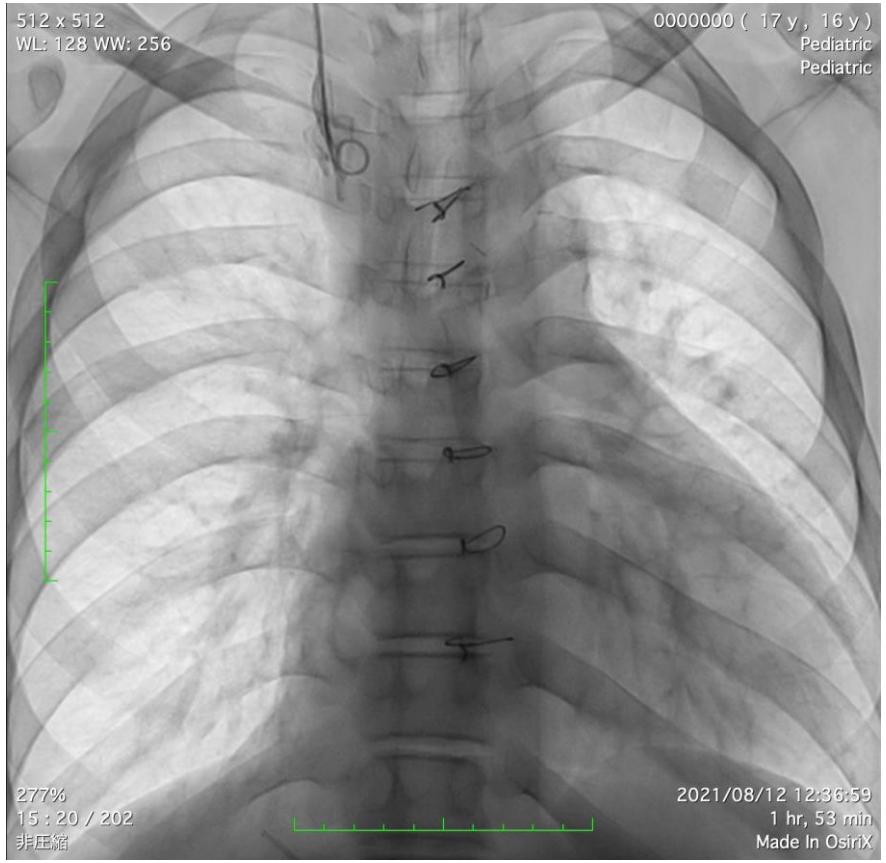
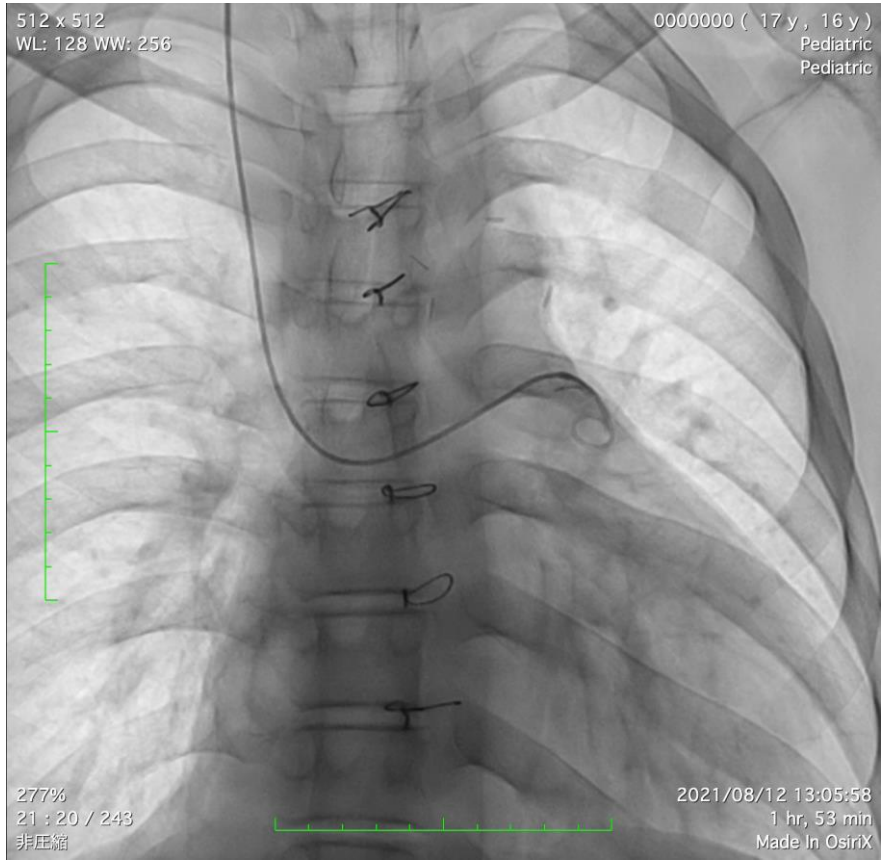
Preoperative angiogram (SVC, PA), 16 years old





SVC pressure12mmHg、 IVC pressure 7mmHg、

SVC→collateral vein→PA→SVC Failed Glenn circulation

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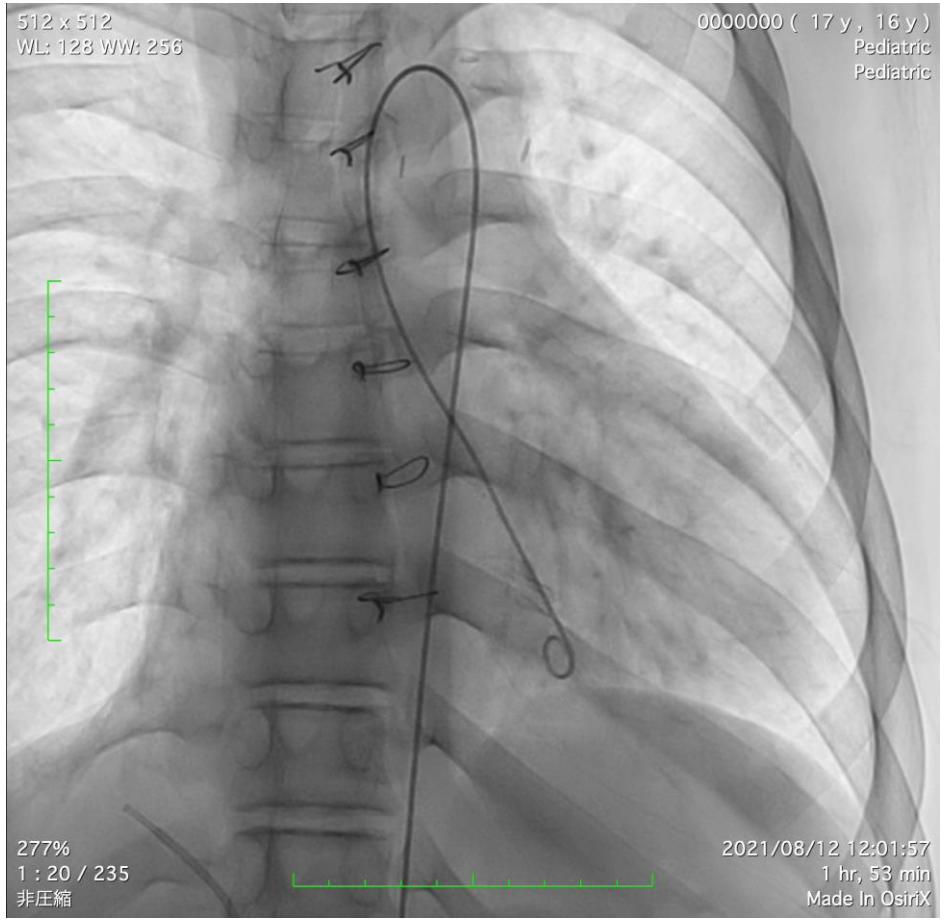
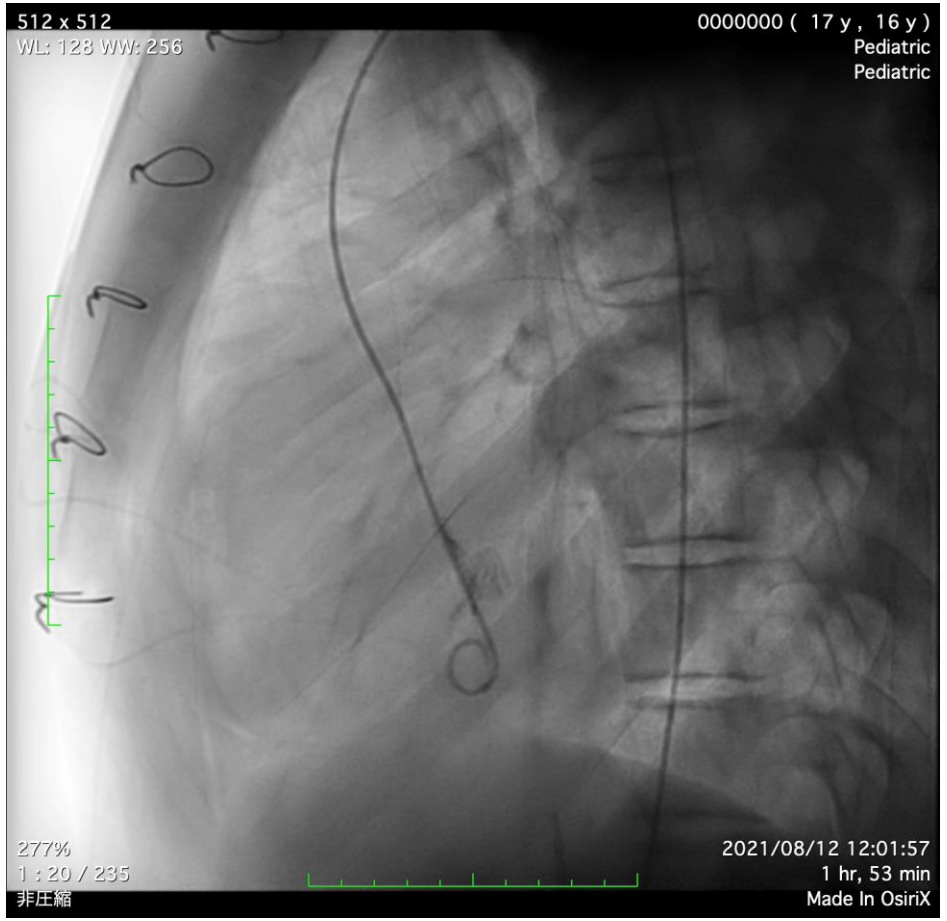
Preoperative angiogram(LV), 16 years old





LVEDV220% of normal、 LVEF51%

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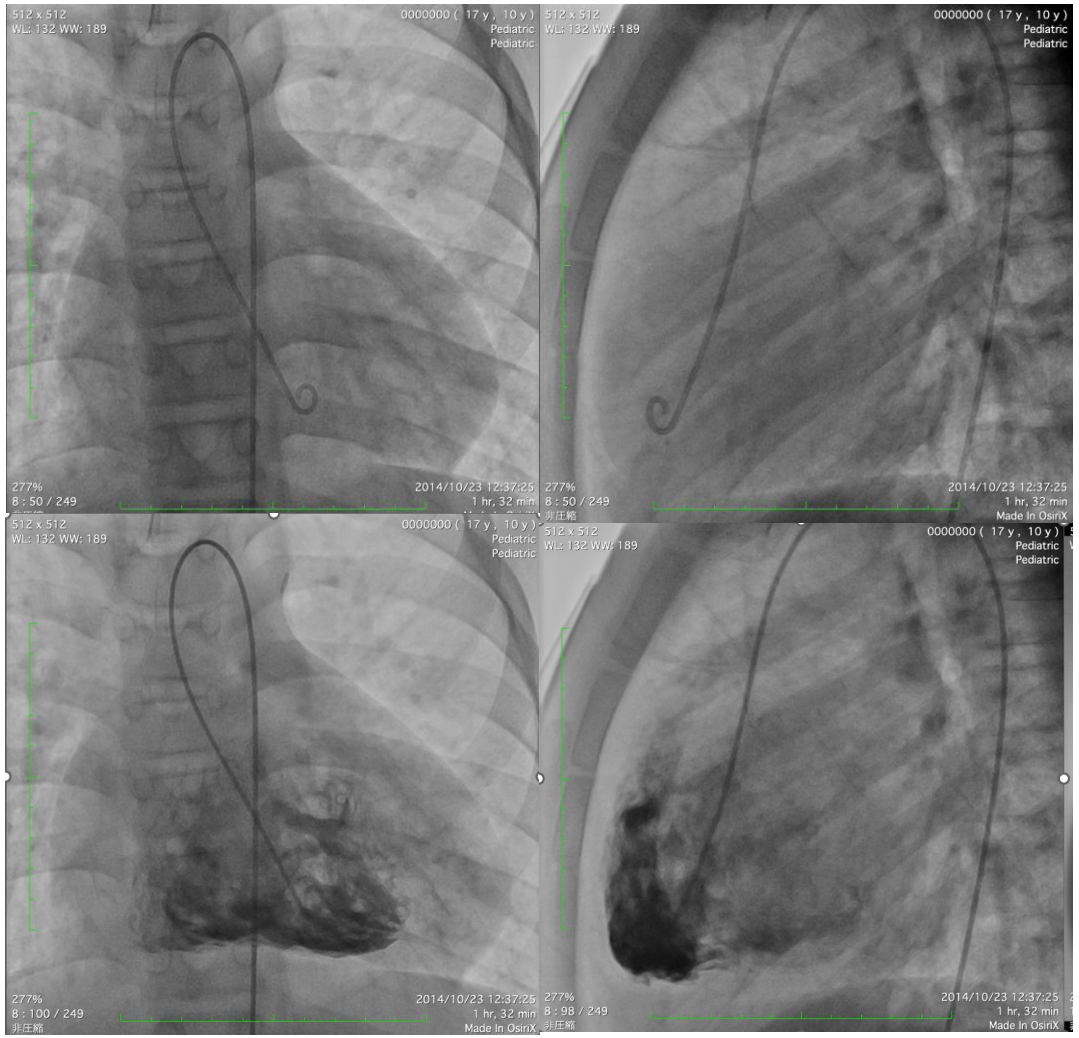
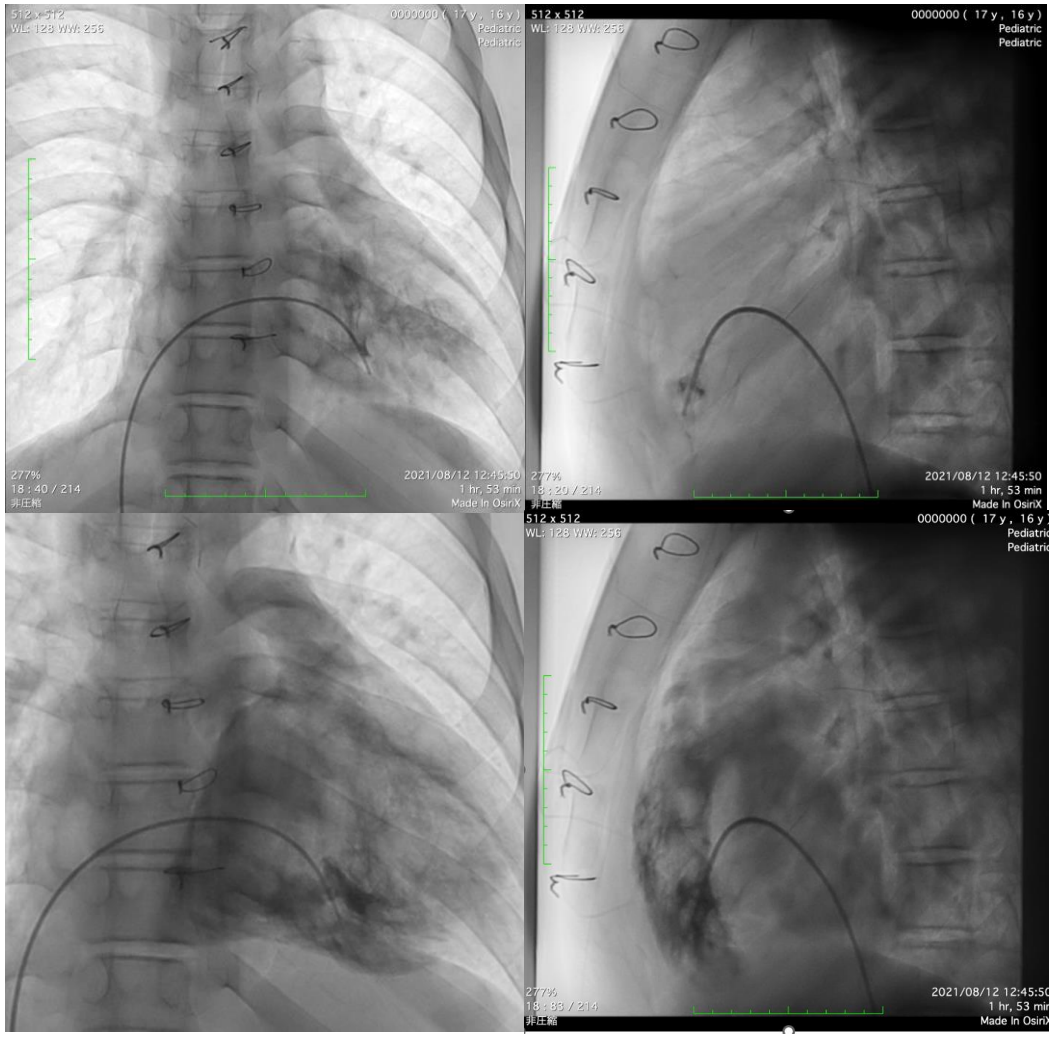
RV angiogram （10 years old vs. 16 years old



10 years old　RVEDV 29%　of normal



16 years old　RVEDV 113% of normal

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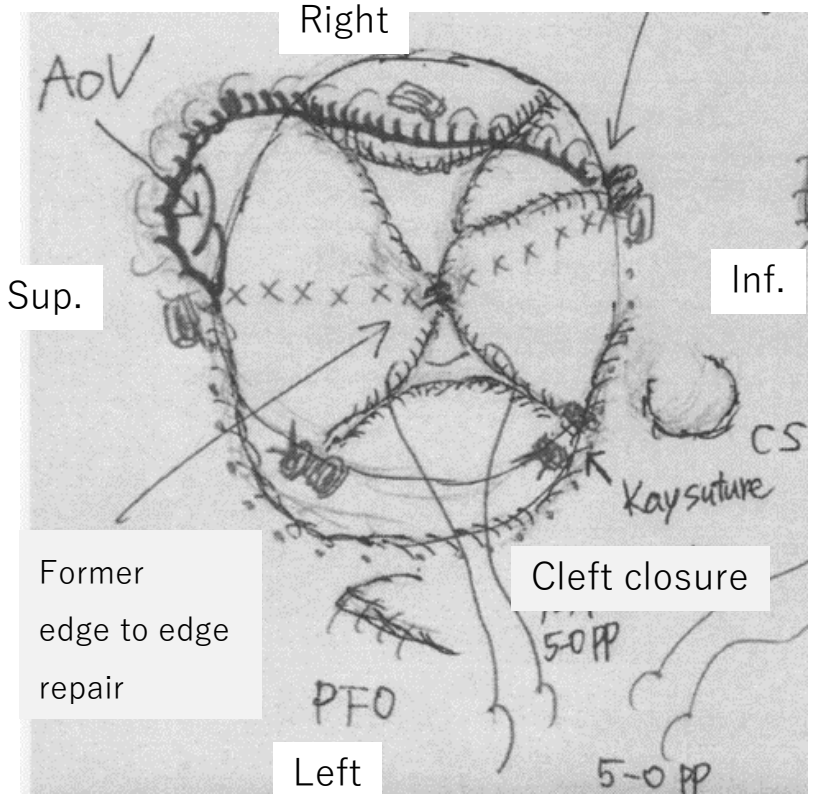
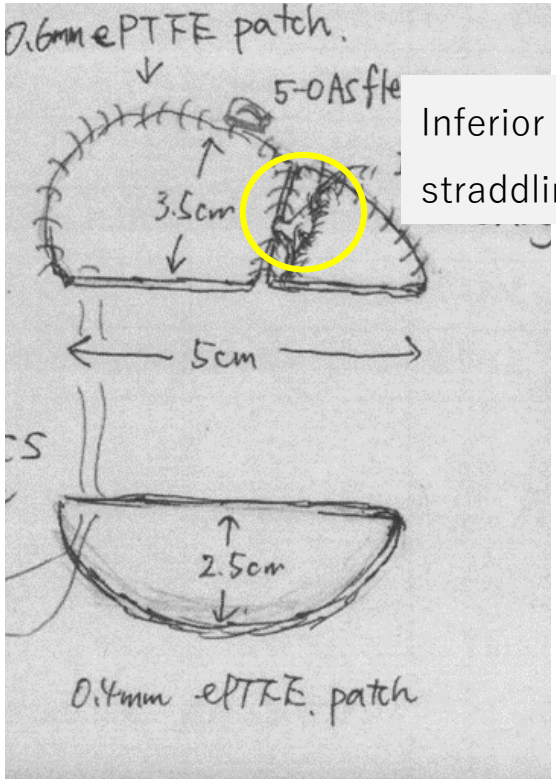
Intraoperative Findings & Surgical Repair 1





Inferior bridging leaflet chorda straddling

VSD patch separation
Tear-drop shape VSD patch
Patch height 3.5cm

Semilunar shape ASD patch

Patch height 2.5cm

- Left side cleft complete closure
- S-LL, I-LL commissuroplasty
- Rt. AVV size 88 % N (ϕ 25/28.3)

- VSD・ASD patch closure
- VSD patch separation and re-suture to preserve a straddling chorda of inferior bridging leaflet

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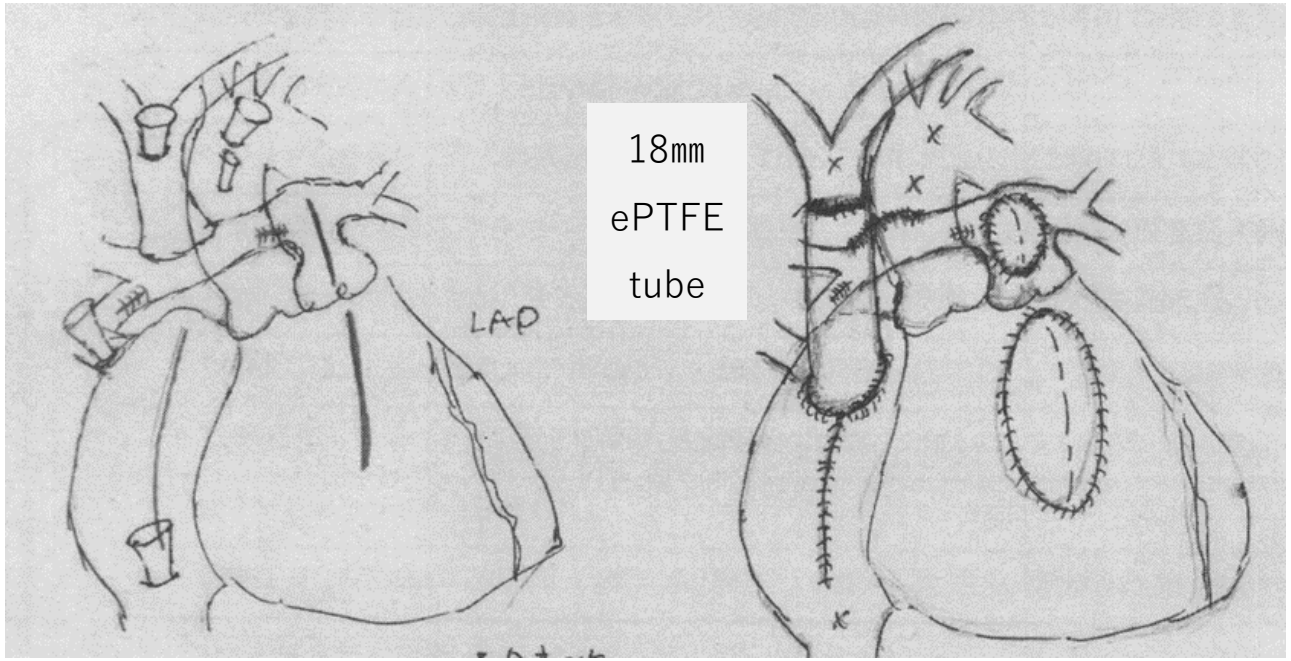
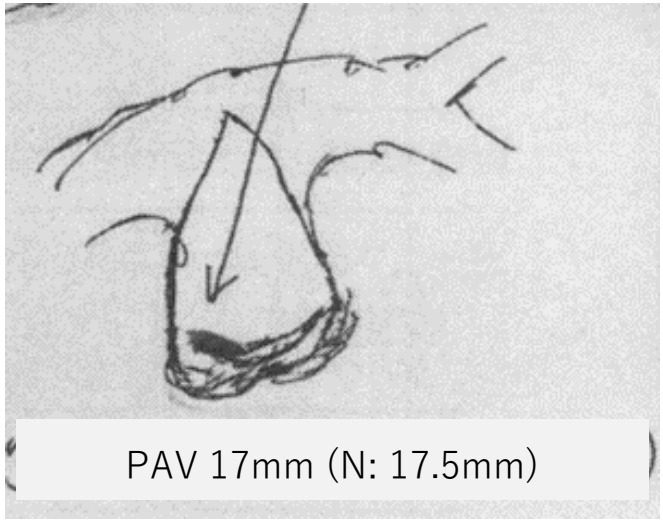
Intraoperative Findings & Surgical Repair 2





PAV 17mm (N: 17.5mm)

- Bidirectional Glenn anastomosis→SVC reconstruction with 18㎜ ePTFE tube
- Pulmonary valve commissurotomy, main PA plasty, RVOT muscle resection.
- Cardiac arrest time 186 minutes. CPB time 463 minutes.

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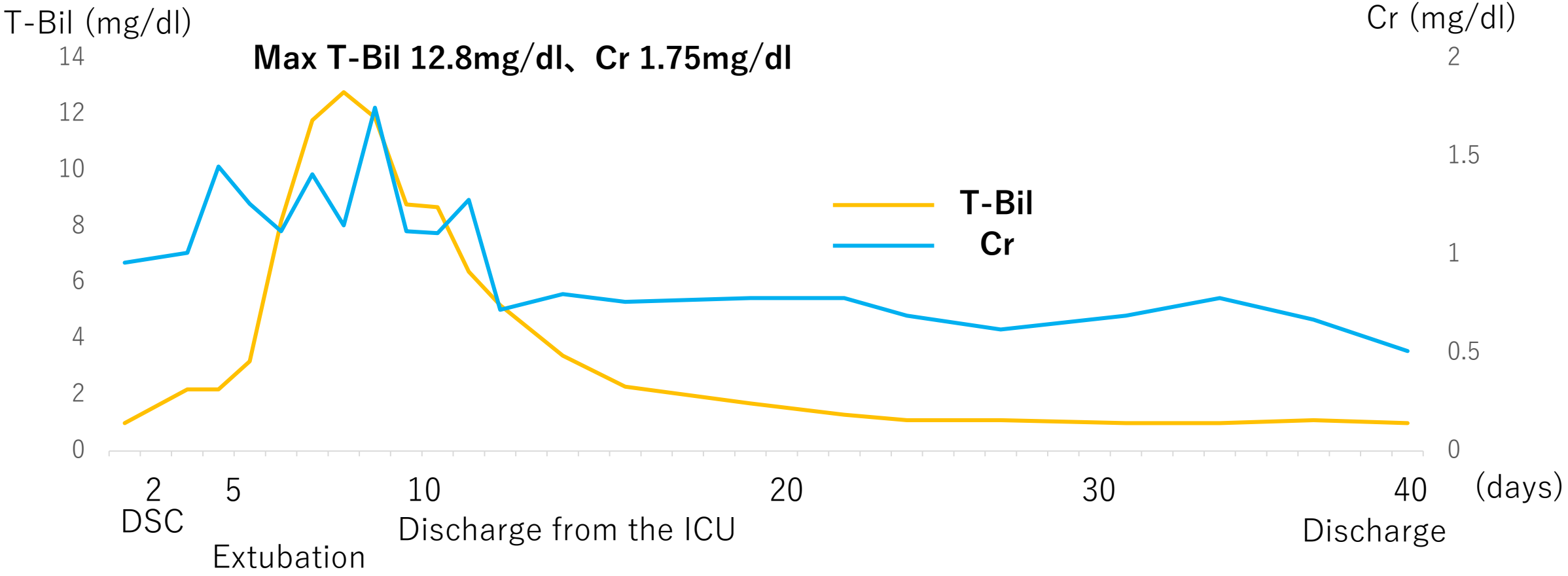
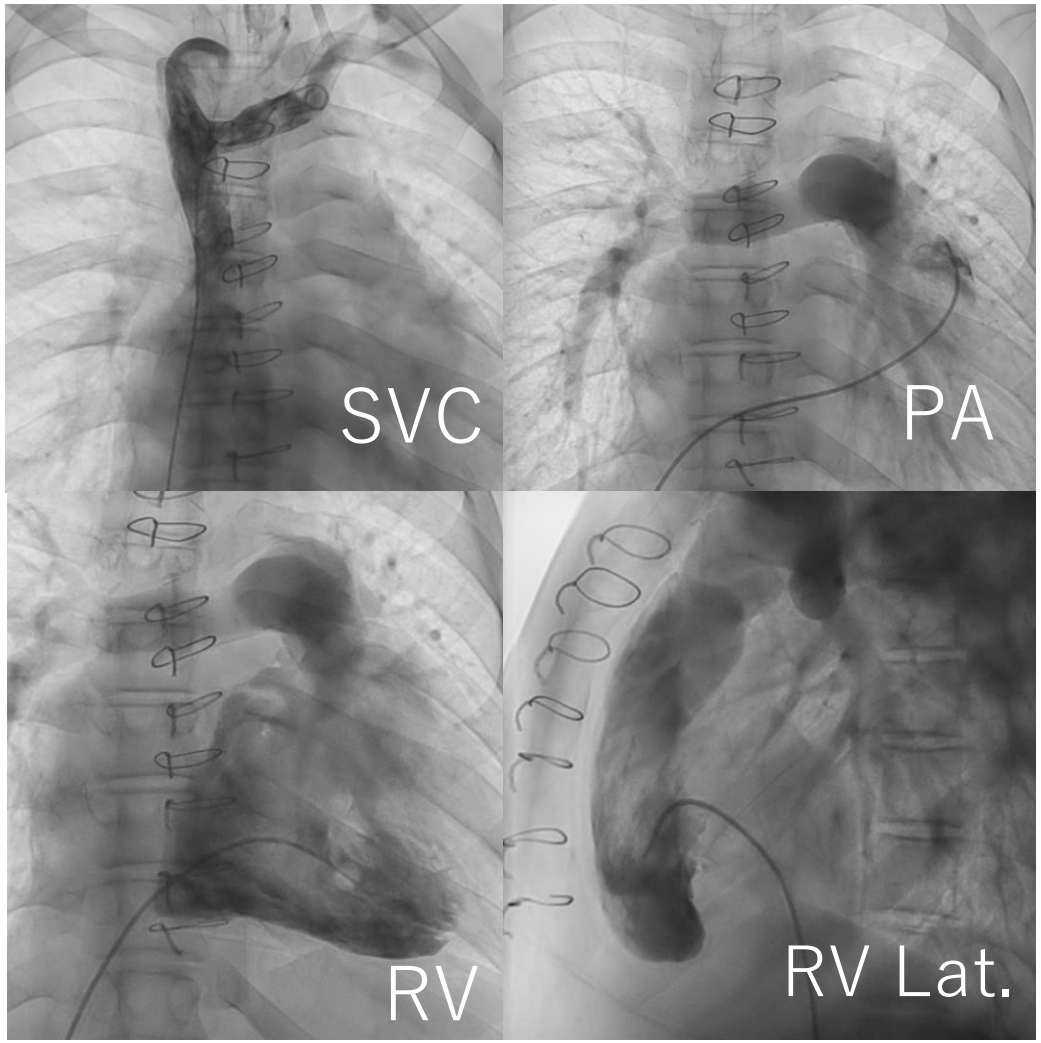
Postoperative course



The patient was discharged on postoperative day 40 under administration of pulmonary vasodilators without hepatic or renal impairment. (SpO_2 98%, O_2 1L/minute)

Postoperative angiogram (14 months)



SVC=IVC=RA (7)

RV 35/8 RVEDV 97%
PA 21/10(17)
PCW 13

LV 84/12 LVEDV 220%
Ao 82/53 (mmHg)

Rt. AVVR 2°
Lt. AVVR 1°

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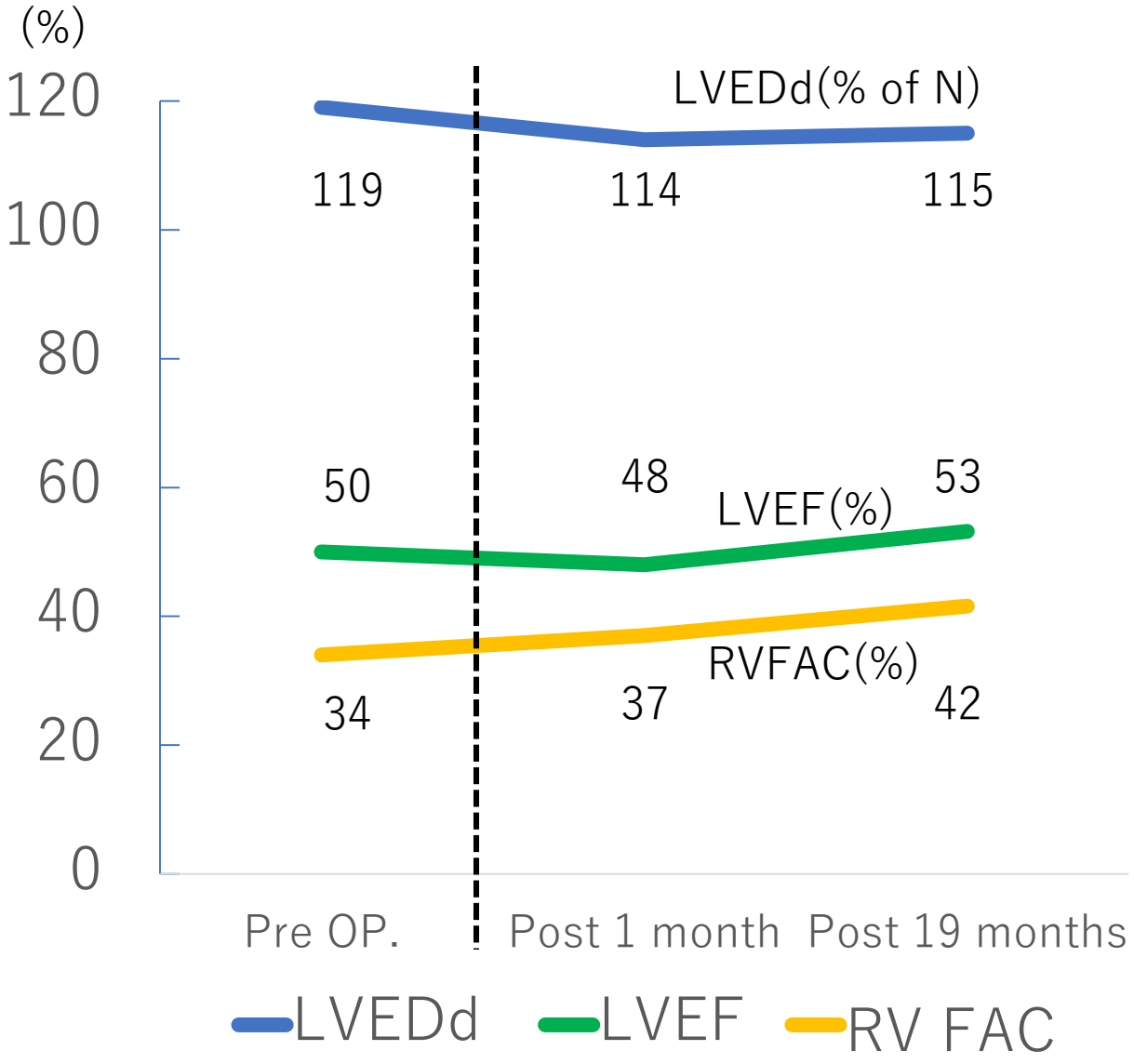
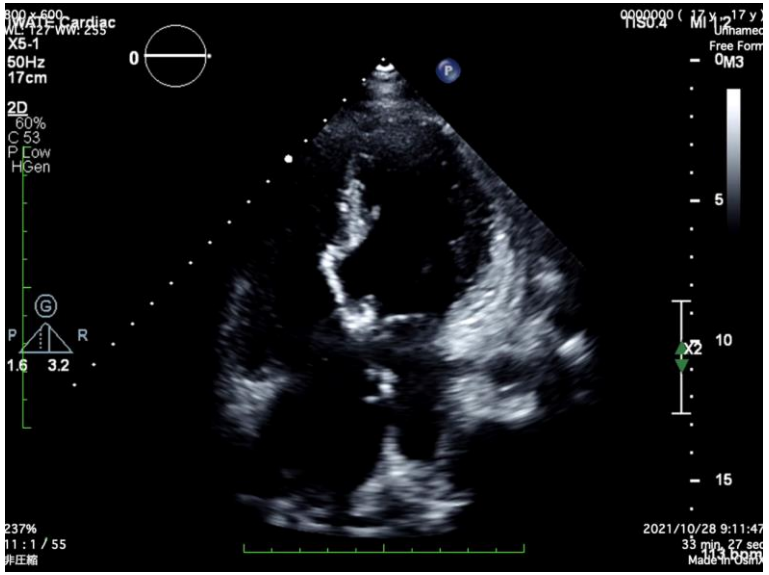
Post operative echocardiogram(19 months)





LVEDd 115% of Normal
LVEF 53 %

RV FAC　42 %

MR mild, TR mild, LVOTS(-),
RVOTS（±）2.2m/s

Discussion

The trends of ventricular volume (normal ratio)

	10 years old, 28kg	16years old, 50kg
RVEDV (ml)	19ml (29%)	126ml (113%)
LVEDV (ml)	156ml(226%)	251ml (220%)

Factors of RV enlargement

: RVOT muscle resection (10 years old)

Volume overload due to failed Glenn circulation & CAVV regurgitation

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Conclusion

- This case represents a rare example that long-term ventricular volume overload led to ventricular enlargement, enabling biventricular repair.

- This course suggests an importance of re-evaluation and a possibility of ventricular functional improvement with subsequent biventricular conversion.

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