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1 RESEARCH LETTER

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3 **Tetralogy of Fallot with coronary artery to pulmonary artery fistula**

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5 Running head: Tetralogy of Fallot -coronary to artery fistula

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25 The authors declare no conflict of interest

1   WHAT'S ALREADY KNOWN ABOUT THIS TOPIC?

- 2   •   Coronary artery fistulas (CAF) are rare anomalies with an incidence of 0.002 % in
3   the population.

4

5   WHAT DOES THIS STUDY ADD?

- 6   •   The association of tetralogy of Fallot, CAF and agenesis of the ductus arteriosus
7   has not been reported

- 8   •   The discrepancy between the narrow pulmonary annulus with thickened
9   pulmonary valve and the large pulmonary trunk can be caused by extra blood
10   supply through the large fistula.

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1 We report a right coronary artery to pulmonary artery fistula as a rare congenital
2 malformation in a patient with tetralogy of Fallot and absent ductus arteriosus.
3 A healthy 33-year-old woman, gravida 3, para 1, was referred at 18+4 weeks with
4 suspicion of a fetal cardiac malformation. Her brother died neonatally from a
5 transposition of the great vessels.
6 Ultrasound examination in this pregnancy revealed a tetralogy of Fallot (TOF) with a
7 right-sided aorta and a narrow pulmonary artery annulus with only mild antegrade
8 flow, while the pulmonary trunk was well developed (Figure 1). No extracardiac
9 anomalies were seen. An amniocentesis showed a normal male CNV-profile.
10 The couple decided to terminate the pregnancy at 23+5 weeks of gestation. The
11 autopsy showed a TOF with a right-sided aortic arch and agenesis of the ductus
12 arteriosus. Remarkable is the wide origo of the right coronary artery with a larger
13 artery branching from this right coronary artery and passing ventrally of the
14 pulmonary truncus and draining above the left anterior cusp into the lumen of the
15 pulmonary truncus (Figure 2). The left coronary artery was normal and no
16 extracardiac anomalies were found.
17 Congenital coronary artery fistulas (CAF) are characterized by a normal aortic origin
18 of the coronary artery involved but with a fistulous communication with the atria, the
19 ventricles or with the pulmonary artery, coronary sinus or the superior vena cava.
20 Coronary artery fistulas are rare anomalies encountered in 0.2% of angiographic
21 series. The incidence in the overall population is estimated 0.002 %. The right
22 coronary artery is most often involved in clinically detectable CAF. Fistulas most
23 frequently involve the right ventricle, closely followed, in terms of incidence, by
24 drainage into the right atrium or pulmonary artery. Left coronary fistulas are less
25 common, but usually drain into the right ventricle or right atrium. While coronary-

1   cameral fistulas are abnormal connections between a coronary artery and a cardiac
2   chamber, the embryological basis of coronary artery to pulmonary artery fistulas can
3   be based on Hackensellner's involution persistence hypothesis¹ proposing that there
4   are six anlagen in the truncus of which two, that are seen in the aortic sinuses, persist
5   and give rise to the coronary arteries while the others involute. Accordingly, the
6   normally involuted anlage from the pulmonary sinus persists and connects with the
7   anlage from the aortic sinus giving rise to these fistulas.² Small congenital CAF can
8   be asymptomatic, larger more complicated CAFs can cause congestive heart failure,
9   myocardial ischemia due to a coronary steal effect, thrombosis, rupture and infective
10   endocarditis. Symptomatic CAF can be treated by selective ligation of the fistula tract
11   or transcatheter closure.

12   CAF can be found in isolation, or as in the present case in association with other
13   structural heart disease. The postnatal diagnosis can be suspected by
14   echocardiography, but cardiac catheterization is required to precisely evaluate the
15   vascular anatomy³. Isolated giant CAF has been diagnosed at 38 weeks of gestation,
16   and successfully treated in the neonatal period⁴. The prenatal diagnosis of ventriculo-
17   coronary artery fistula in the setting of PA-IVS has also been made previously⁵. In the
18   literature coronary-pulmonary artery fistulas (CPAF) have been reported in
19   association with TOF and pulmonary atresia in 10 % and the majority of them have a
20   fistula originating from the left coronary artery with >50 % also having systemic-to-
21   pulmonary collateral arteries (MAPCAs)⁶.

22   Only one report described the prenatal diagnosis of a large fistula from the left
23   coronary artery to the left atrial appendage in TOF, with congestive cardiac failure
24   postnatally and death after surgical correction of TOF with ligation of the fistula at 3
25   months of age⁷.

We did not find the association of TOF , CPAF and agenesis of the ductus arteriosus as described in the present case. The fistula was only diagnosed at autopsy , but prenatally we found a discrepancy between the narrow pulmonary annulus with a thickened pulmonary valve with minimal flow at 22 weeks and the pulmonary trunk that was adequately developed. This adequate pulmonary development can be caused by extra blood supply through the large fistula.

ETHICS

Approval of the ethical commission was not required for this manuscript.

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1 **Legend to the figures**

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3 **Figure 1** Ultrasound with white pulmonary valve (arrow) , well developed trunk

4 (dot) and right sided aorta.

5

6 **Figure 2** Macroscopic view on the heart with aorta, pulmonary artery,

7 right coronary artery and the fistula.