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Management of isolated right aortic arch with prenatal diagnosis in two French M3 C centers

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Background Prenatal diagnosis of isolated (i.e. without intracardiac anomaly) right aortic arch (IRAA) increases [1], mostly due to the integration of the tracheal-vessel image in fetal screening [2]. IRAA may constitute an encircling anomaly with functional digestive or respiratory repercussion [3,4], and is sometimes associated with genetic anomalies [5], especially the microdeletion 22q1.1 [6—8]. However, antenatal and postnatal cares are not consensual and based only on partial data. The aim of this study was to describe the management of this new kind of population in two French M3 C centers.

Method We conducted a retrospective observational study in two M3 C competence centers (Montpellier and Tours) from 2010 to 2018 including all patients with a prenatal diagnosis of IRAA.

Results We included 36 patients and observed an increased prenatal diagnosis among time in both centers (Fig. 1). Amniocentesis were performed in 27 cases (75%) and conducted to the diagnosis of del22q1.1 in one case (3%). After birth, 16 children were diagnosed for encircling anomaly, but only 4 (11%) needed a surgical repair, all for a double aortic arch. Comparing the practice in the two centers, we found an increased number of tomodensitometry (67% versus 27%; $P = 0.04$) and proportionally an increased number of diagnosis of encircling anomalies (62% versus 14%; $P = 0.02$) in center 1 (Table 1). Indeed the scanner, when performed, completed the echocardiographic prenatal diagnosis in 100% of cases.

Conclusions The lack of harmonization of practices concerning the prenatal diagnosis of IRAA may have consequences for patients care. Therefore, we begin a national multicentric retrospective study between 2010 to 2019 to collect data about the management of prenatal diagnosis of IRAA: ARCADE (2019_IRB-MTP_03-31) that aims to evaluate the reliability of antenatal diagnosis of IRAA in terms of anatomy and functional prognosis to harmonize the medical care for this population.

Keywords Isolated right aortic arch; Prenatal diagnosis; Pediatrics; Congenital heart disease; Del 22q1.1

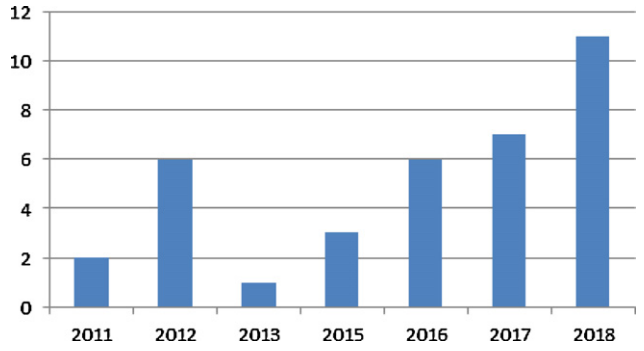

Fig. 1 Increased incidence of prenatal diagnosis of isolated right aortic arch in both M3 C competence centers.

Table 1 Main results of the pilot study in two M3 C competence centers.

	Center 1: Montpellier $N = 21$	Center 2: Tours $N = 15$	P value
Number of amniocentesis	15 (70%)	12 (80%)	0.71
Complication of amniocentesis	0	0	1
Micro del 22q1.1	0	1 (7)	0.42
Symptomatic children	6 (28%)	4 (27%)	1
Number of tomodensitometry	14 (67%)	4 (27%)	0.04
Number of encircling anomalies	13 (62%)	3 (14%)	0.02
Double arch	2 (9%)	2 (13%)	1
Surgical repair	2 (9%)	2 (13%)	1

Disclosure of interest The authors declare that they have no competing interest.

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