Non-cardiovascular findings on chest CT angiography in children with congenital heart disease: How important are they?

Yaotse Elikplim Nordjoe*†, Suzanne Rita Aubin Igombe† and Latifa Chat†

Abstract
Background: There are only a few publications about the non-cardiovascular findings in children with congenital heart diseases explored by chest CT angiography. The purpose of our study is to evaluate the prevalence of non-cardiovascular findings on chest CT angiographies in children with congenital heart disease and to raise awareness about their importance among the radiologists.

Methods: We retrospectively reviewed the 272 chest CT angiographies performed in our pediatric radiology department between January 2017 and March 2021 and extracted the data of the 180 patients positive for a congenital heart disease. Then from that pool, we sorted out the non-cardiovascular findings into significant and non-significant in regard of their relevance or not in the patient’s management.

Results: Non-cardiovascular lesions were found in 58% (105/180) of our patients, and 49% (88/180) of them presented at least one significant non-cardiac lesion. Lung and airways abnormalities were found in 41% (74/180) of the cases, representing the majority of the non-cardiovascular findings. Syndromic associations were found in 17% (28/180) of our patients, including 14 cases of heterotaxic syndrome.

Conclusion: Non-cardiovascular findings are common in children with congenital heart disease. Reporting these associated lesions is a requisite for an optimal therapeutic management of these children. Radiologists should never forget that a Chest CT angiography in children is first and foremost a chest CT.

Keywords: Congenital heart disease, Chest CT angiography, Children, Non-cardiovascular findings

Background
The congenital heart diseases are relatively common and very diverse, with an estimated prevalence of 1–4 per 1000 births [1–3]. In children, in order to align with the ALARA “As Low As Reasonably Achievable” principle, cardiac ultrasound and MRI represent excellent imaging tools for diagnosing and monitoring patients with congenital heart disease [4, 5]. However, they are not perfect and not suitable for all situations.

With the development of multi-detector scanners, dose reduction techniques, better reconstruction algorithms and global advances in the field of computer science, chest CT angiography has become and accepted as a rapid and non-invasive diagnosing and monitoring tool in children with congenital heart disease [4, 6–11].

CT allows a detailed regional analysis and therefore, enables the detection of findings other than cardiovascular. These non-cardiovascular findings can have a major impact in the management of the patient. Their prevalence and implications have been the subject of

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numerous publications in adult cardiac pathology with a highly variable prevalence ranging from 7.1 to 81% [12–19]. On the contrary, in the pediatric population, there are only three publications studying non-cardiovascular findings in congenital heart disease, two on CT angiography [20, 21] and the other on MR angiography [22].

The aims of our work are to evaluate the prevalence and relevance of non-cardiovascular findings on chest CT angiography in children with congenital heart disease in our institution and to raise awareness among the radiologists about the importance of their detection.

**Material and methods**

**Description of the study**

This is a retrospective observational study over a period of 4 years and 3 months (between January 2017 and March 2021). Analysis was performed on the database of the radiology department of the children’s hospital in Rabat—Morocco. We reviewed all the 272 chest CT angiographies performed in the above-mentioned period, for a known or suspected congenital heart disease, of which the 180 patients positive for a congenital heart disease formed the pool of our study. From that pool, non-cardiovascular findings have been identified and processed to evaluate their prevalence and their relevance.

Patients had to be less than 18 years old.

Since the study is based on a retrospective analyze of our archiving database, the obtention of a written informed consent was waived by our institution ethical committee.

**CT angiography procedure**

**Preparation of the patients**

A reassurance of the patient was achieved by the parents and the medical staff. If needed, one of the parents was authorized—after protection—to remain in the machine room with the child. A peripheral intravenous access was obtained using a 20 to 24 Gauge catheter placed in an antecubital forearm or hand vein. A little sedation was sometimes necessary; performed by the anesthesiologist assigned to the scanner.

**CT examination protocol**

All the exams were performed on a single-source 16-channel MDCT scanner (SIEMENS Somatom Emotion 16).

An integrated pre-set chest CT angiography protocol was used, allowing the adaptation of the acquisition parameters to the pediatric population: 80 - 100 kV, with for example 30 mAs at 3 kg and 40 mAs at 5 kg. All the examinations were non-electrocardiogram-gated and were acquired during shallow free-breathing or suspended inspiration. Contrast medium was power injected (Medrad; Stellant), using 1–2 ml/kg of nonionic contrast medium (Ultravist 300 mg I/ml; Bayer Schering Pharma) at a rate of 0.5–3.0 ml/s, followed by 10–30 ml of saline at a similar rate. Automated bolus tracking was used to achieve optimal opacification. Data sets were reconstructed into 1 to 1.5 mm contiguous slices. We only used filtered back projection algorithm for the reconstructions. Reconstructed slice thickness was 1.5 mm with H30 kernel for mediastinum and 1 mm with H80 kernel for lung. The mean DLP was 45.56 mGy/cm (±23.81). The mean CTDIvol was 1.57 mGy (±0.52).

**Definition of non-cardiovascular findings**

Non-cardiovascular findings are abnormalities other than those of the heart and vessels, discovered on the volume acquired.

A non-cardiovascular finding was “significant” if further investigations were needed and/or if this abnormality contributed to the worsening of the patient’s clinical state.

A “significant” finding could either be “expected” or “unexpected”. An “expected” lesion is one whose occurrence is not surprising, being part of the usual associated lesions of that particular congenital heart disease while an “unexpected” lesion is just the opposite of that.

It is important to note that the same finding can be significant in one patient but non-significant in another one, depending on its impact on the patient’s condition. Also, there are some findings whose categorization cannot be formally certain because the physio-pathological implications are complex.

**Data collection and analysis**

Post processing and analysis were carried out on a dedicated post processing workstation with SyngoVia software, by a certified senior radiologist with 10 years of experience in congenital heart diseases (Dr L.C.).

The entire arsenal of this platform was used, allowing both usual multiplanar reformattting and MIP, MInIP, VRT 3D reconstructions etc.

Images and reports were archived on our picture archiving and communication systems VISIONPACS and VISIONRIS.

The retrospective analysis of the data was carried out on these above-mentioned archiving platforms. First, all CT scans performed for the indication of known or suspected congenital heart disease were extracted. A second sorting allowed the separation of the normal examinations from the examinations on which there was indeed a congenital heart disease. Lastly, the pool of congenital heart disease was analyzed to deduce those with associated non-cardiovascular findings. These non-cardiovascular findings were subcategorized into: infectious,
malformative, tumoral lesions and others. Also, an anatomi
cal distribution of these abnormalities was made, into supra-diaphragmatic findings (pulmonary and air-
ways, mediastinal) and infra-diaphragmatic findings
(liver, kidney, spleen, bowel). Bone skeletal abnormalities
have also been isolated and finally, syndromic associa-
tions were identified.

Results
Overall results
A total of 272 chest CT angiographies was performed for
a known or suspected congenital heart disease, of which,
180 was positive for a congenital heart disease. The mean
age was 3.1 years (± 3.2), ranging from 1 day to 17 years.
Some of our patients (n = 20) had a history of surgery
(Table 1).

We detected 155 non-cardiovascular anomalies in
105 patients positive for congenital heart disease (58% of
180), of which 88 patients had significant anomalies
and 17 patients had non-significant anomalies. More
than one non-cardiovascular anomalies were found in
24 patients (23% of 105). In these cases where there were
association of several abnormalities in the same patient,
if there is only one significant finding; the patient was
classified as significant (Table 2).

Non-cardiovascular findings were classified by sub-
categories: infectious, malformative, tumoral and others
(Fig. 1).

| Table 1 | Characteristics of the study population |
|-----------------|----------------------------------------|
| Population with congenital heart disease | Mean age ± SD | Range |
| n = 180 (99 M / 81 F) | 3.1 years ± 3.2 | 1 day–17 years |
| Patients with or without non-cardiovascular findings | n = patients |
| Patients with non-cardiovascular finding | 105 (58% of 180) |
| Patient without non-cardiovascular findings | 75 (42% of 180) |
| History of surgical procedure | n = patients |
| Blalock-Taussig-Thomas | 11 |
| Coarctation repair | 3 |
| Fontan | 1 |
| Pulmonary artery prosthesis | 1 |
| Tetralogy of Fallot repair | 3 |
| Senning | 1 |
| Repartition by congenital heart disease | n = patients |
| Left obstructive lesions | 42 |
| Right obstructive lesions | 33 |
| Cono-truncus anomalies | 30 |
| Congenital arch anomalies | 30 |
| Septation anomalies | 35 |
| Pulmonary venous anomalies | 10 |

| Table 2 | Distribution of significant and non-significant findings |
|-----------------|----------------------------------------|
| Non-cardiovascular findings | n = Patients (%) of 180 |
| Significant | 88 (49%) |
| Expected | 66 (37%) |
| Unexpected | 22 (12%) |
| Non-significant | 17 (9%) |
| Total | 105 (58%) |

Topographic distribution and syndromic associations
The non-cardiovascular findings were subdivided into
four main groups: supra-diaphragmatic, infra dia-
phragmatic, skeletal and syndromic associations.

Supra-diaphragmatic anomalies (Table 3)
Two subsets were isolated. On the one hand lung and
airways findings were identified in 74 (41% of the
180 patients of our series) patients and on the other,
mediastinal findings were identified in 3 patients. Lung
and airways findings were by far the most frequently
encountered anomalies, mainly dominated by infec-
tious pneumopathies, mosaic attenuation pattern and
atelectasis. Associations can be found in the same
patient.
Infra-diaphragmatic anomalies (Table 4, Figs. 2, 3)
Four subsets were isolated: Liver, kidney, spleen and bowel. Splenic anomalies were essentially asplenia or polysplenia mainly integrated into heterotaxic syndromes. The most frequent liver abnormalities are represented by hepatic congestion, due to hemodynamic changes. Bowel anomalies were essentially malformative with a predominance of hiatal hernias. Kidney anomalies were also malformative, with a horseshoe kidney being the predominant finding.

Skeletal anomalies (Table 4; Fig. 4)
Since the CT coverage excluded the limbs, skeletal findings were solely represented by spine anomalies found in 5 patients.
A total of 28 cases of syndromic association were identified, representing approximately 16% of the patients presenting a congenital heart disease and approximately 27% of the patients (28 of 105) positive for a non-cardiovascular CT finding. Heterotaxic syndromes were predominant.

Discussion

The prevalence of non-cardiovascular anomalies is 58% in our study. In similar studies using CT angiography as the exploration technique, this prevalence is 83% in the series by Malik et al. [21] and 31.6% in the series by Rodríguez Martin et al. [20].

There is a moderate variability between these different series. Also, despite the use of a 64 detectors MDCT scanner, Malik et al. [21] and Rodríguez Martin et al. [20] showed a significant difference in the prevalence of the non-cardiovascular findings.

In view of these observations, we deduce a few reflections: we used a 16-detectors MDCT scanner. It is clear that a 64-detectors machine is more efficient. Also, it would be justified to conclude that the higher prevalence of Malik et al. [21], compared to our study, is in a large part, due to the use of a more efficient machine. But this reasoning fails when we compare the results of Malik et al. [21] with those of Rodríguez Martin et al. [20]. This could mean that the performance of the MDCT scanner and the characteristics of the population are both, the key factors determining the prevalence of non-cardiovascular findings. Because the study by Malik et al. [21] was carried out in a reference center specialized in pediatric vascular imaging; therefore, recruiting patients in greater number and severity; whereas our study and that of Rodríguez Martin et al. [20] were carried out in a pediatric imaging department with several activities, some of which are dedicated to pediatric cardiovascular imaging.

Nevertheless, it is obvious that non-cardiovascular findings are frequent. A common point of these three studies is the higher frequency of pulmonary and airway findings observed in 41% of patients, versus 59% in Malik et al. series [21] and 16.7% in that of Rodríguez Martin et al. [20].

Ghadimi Mahani et al. [22] were interested in the prevalence of extra-cardiac abnormalities found on MR angiographies in children with congenital heart disease. They found an overall prevalence of 16.5% of non-cardiac anomalies with a predominance of abdominal findings (51% versus 31% for intrathoracic findings). This difference can be explained by the lower performance of MRI for the study of thoracic structures.
These above-mentioned studies are listed along with ours in (Table 6) showing comparison of their key points. The importance of these findings varies. Some of them are minor, without significant clinical consequence; others are major, requiring additional investigations or even appropriate immediate treatment. In our series, 49% of patients with congenital heart disease have at least one significant non-cardiac finding, requiring additional investigations or specific immediate or deferred management. This rate was 74% in the series of Malik et al. [21], higher than in ours. This high prevalence of significant non-cardiovascular findings detected on examinations performed for congenital heart disease, requires the radiologist to be vigilant and attentive when analyzing a chest CT angiography in children. The impact of associated non-cardiovascular lesions on the diagnostic and therapeutic management of the patient can be crucial [19, 21–24]. However, some authors believe that the detection of non-cardiovascular anomalies on cardiac CT angiographies (especially in adults) can be rather a disadvantage, because their detection often leads to additional diagnostic procedures and additional costs without significant improvement of the patient’s condition [25, 26].

Such reasoning goes against the goal of our study and many other similar ones, because it could cultivate the laziness of the radiologists as regards to the systematic analysis of all the volume acquired. We also believe that ignoring or not reporting such anomalies is a mistake and could have a negative impact on the patient’s outcome.

As a reminder, the data of our study shows that approximately one out of two patients with congenital heart disease have at least one significant non-cardiac finding. Moreover, these significant anomalies can be unexpected, urging the radiologists to look at everything and to report the other findings associated with the congenital heart diseases. The vast majority of the congenital heart diseases are isolated. However, 20–30% are the expression of a genetic abnormality fitting into a syndromic framework. Genetic progress has been made in identifying several associated genes in polymalformative genetic syndromes that combine congenital heart disease and extra-cardiac manifestations [27–30]. The knowledge of the
syndromic character of a heart disease requires a systematic investigation of commonly associated extracardiac abnormalities.

In our series, we identified 28 cases of syndromic heart diseases, representing 17% of all our patients and 27% of patients with non-cardiovascular findings. These syndromic heart diseases have a prevalence of 22% in the series of Malik et al. [21] among which half had genetic confirmation only after radiological suspicion. This again underlines the importance of detecting and reporting these findings.

Critical analysis of the study

The first bias is the retrospective nature of the study. There is an intrinsic incomplete nature of the recruitment of patients and of the data collection because, for example, if a desired data is missing from the report, it is considered to be really absent when it could have been present if the study had been prospective and we had systematically planned to search for particular anomalies.

Also, given the retrospective nature of our study, there are chances of lack of adequate monitoring of the evolution of non-cardiovascular abnormalities in order to effectively weigh their importance in the management of the patient.

The data collected depends on the skills of a single reader. It is obvious that adding a second one would minimize the risk of omission and add more value to our observations. Despite this, we are confident in our excellent diagnostic ability in congenital heart diseases.

The separation between significant and non-significant findings can sometimes be delicate. In addition, a direct attribution of an anatomical abnormality to a clinical manifestation is not obvious because the pathophysiological implications can be complex.

Table 5 Distribution of syndromic congenital heart diseases

| Syndromes            | n = Patients |
|----------------------|-------------|
| Heterotaxic syndromes | 14          |
| Down Syndrome        | 4           |
| Williams Beuren      | 3           |
| Kartagener           | 3           |
| 22q11.2 (Di-George)  | 2           |
| VACTERL              | 1           |
| Marfan               | 1           |
Last but not least, we used a single source 16-detectors MDCT scanner. It obvious that a better machine would have grant us a better quality of the data.

Nevertheless, despite these limitations, our results are usable. This study allowed us to make a real observation of the prevalence, significance and polymorphism of these non-cardiovascular findings associated with congenital heart disease in children explored by CT angiography.

It should lead us to place more emphasis on the systematic analysis of the acquired volume, in order to detect the maximum associated lesions, since they can have major impact in the management of the patients.

**Conclusion**

Non-cardiovascular findings are frequent on chest CT angiography in children with congenital heart disease. The data of our series showed that almost 2/3 of these children with congenital heart disease have one or several non-cardiovascular anomalies, of which up to 50% can be significant requiring additional investigations and / or contributing to the patient’s symptoms; therefore, requiring immediate or deferred appropriate management. Their detection is essential for the optimal management of these patients.

These observations underline the importance of a systematic analysis of all the acquired volume of chest CT angiographies.
Abbreviations
ALARA: As low as reasonably achievable; MRI: Magnetic resonance imaging; CT: Computed tomography; MIP: Maximum intensity projection; MinIP: Minimum intensity projection; VRT: Volume rendering technic; 3D: Three-dimensional; MDCT: Multidetector-row computed tomography.

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Authors’ contributions
YEN, SRAI and LC equally contributed to this work. YEN: conception, design of the work, acquisition, analysis, interpretation of data, manuscript writing, editing of manuscript. SRAI: conception, design of the work, acquisition, analysis, interpretation of data, manuscript writing, editing of manuscript. LC: conception, design of the work, acquisition, analysis, interpretation of data, manuscript writing, editing and final review of the manuscript. All authors read and approved the final manuscript.

Fig. 6 Kartagener syndrome. Situs inversus (a, b), Bilateral bronchiectasis (c, d)

Table 6 Comparative listing of the different studies

| Study                | Imaging technic | Period                     | Type       | Number of patients (n) | Prevalence of non-cardiovascular findings (%) | Significant non-cardiovascular findings (%) | Predominant findings |
|----------------------|-----------------|----------------------------|------------|------------------------|-----------------------------------------------|---------------------------------------------|----------------------|
| Malik et al. [21]    | CTA             | 2 years and 4 months       | Retrospective | 300                    | 83                                            | 73.7                                        | Lungs                |
| Ghadimi-Mahani et al [22] | MRA             | 4 years and 10 months      | Retrospective | 740                    | 16.5                                          | 2.6                                         | Abdominal            |
| Rodriguez Martin [20] | CTA             | 5 years                    | Retrospective | 222                    | 31.6                                          | 22.7                                        | Lungs                |
| Our study            | CTA             | 4 years and 3 months       | Retrospective | 180                    | 58                                            | 49                                          | Lungs                |
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Availability of data and materials
The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Declarations

Ethics approval and consent to participate
This study was approved by the local Ethical Committee of our institution (Local Ethical committee of the Children’s Hospital—Centre Hospitalo-Universitaire Ibn Sina of Rabat—Morocco). Consent for publication was waived owing to the retrospective, observational, and anonymous nature of this study. The methods were carried out in accordance with the relevant guidelines and regulations.

Consent for publication
This study was approved by the local Ethical Committee of our institution (Local Ethical committee of the Children’s Hospital—Centre Hospitalo-Universitaire Ibn Sina of Rabat—Morocco).

Competing interests
The authors declare that they have no competing interests.

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