



# Computed Tomography Evaluation of the Paranasal Sinuses in Adults with Primary Ciliary Dyskinesia

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## Abstract

### Keywords

- ▶ aplasia
- ▶ computed tomography scanner
- ▶ Kartagener syndrome
- ▶ paranasal sinuses
- ▶ primary ciliary dyskinesia

**Introduction** Primary ciliary dyskinesia is a rare inherited disease that results in a malfunction of mucociliary clearance and sinonasal complaints. Aplasia/hypoplasia of the frontal and sphenoid sinuses has been described as more frequent in this population. However, to date, no studies have provided a detailed description of computed tomography findings in adult patients with a diagnosis of this condition.

**Objective** To describe the computed tomography (CT) findings of adult patients with primary ciliary dyskinesia.

**Methods** Retrospective observational study of adult patients with primary ciliary dyskinesia who underwent CT.

**Results** Twenty-one adults were included in the study. Aplasia occurred in 38.1% of frontal sinuses and in 14.3% of sphenoid sinuses. Likewise, hypoplasia occurred in 47.6% of the frontal sinuses, in 54.8% of the sphenoid sinuses and in 40.5% of the maxillary sinuses. Furthermore, trabecular loss was identified in 61.9% ethmoidal sinuses. The mean Lund-Mackay score was 13.5. In addition, 9.5% of the patients had concha bullosa, 47.6% had marked bilateral inferior turbinate hypertrophy, 38.1% had marked middle turbinate hypertrophy, and 47.6% had marked septal deviation. Finally, we identified images suggestive of fungus ball, mucocele, osteoma, a possible antrochoanal polyp, and frontal bone erosions.

**Conclusion** The present study provides a detailed description of CT findings in patients with primary ciliary dyskinesia. We also describe abnormalities that must be identified for safer surgical planning and that suggest a diagnosis of primary ciliary dyskinesia if found in patients with a consistent clinical picture.

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## Introduction

Primary ciliary dyskinesia (PCD) is a rare inherited disorder that results in a malfunction of mucociliary clearance and affects 1 in 10,000 to 1 in 40,000 individuals.<sup>1,2</sup> These patients may present with recurrent airway infections, organ laterality defects, and infertility.<sup>3</sup>

These patients often experience early-onset rhinorrhea, recurrent acute rhinosinusitis, or chronic rhinosinusitis (CRS).<sup>4</sup> The prevalence of CRS in the population over 12 years of age has been estimated at 5.51% in the city of São Paulo, Brazil,<sup>5</sup> while in adults with PCD, studies have shown a prevalence of up to 94.8%.<sup>6</sup>

Primary ciliary dyskinesia is as a factor associated with the development of CRS due to ineffective mucociliary clearance.<sup>7</sup> Furthermore, despite conflicting reports in the literature, anatomical changes which can be identified on computed tomography (CT) of the paranasal sinuses have also been identified as possible contributors to the development of CRS. Thus, for example, frontal sinus or ostiomeatal complex drainage could be impaired by the presence of a concha bullosa (CB), paradoxical middle turbinate (PMT), infraorbital ethmoid (Haller) cells, frontoethmoidal cells, suprabullar cells, and supraorbital cells.<sup>8</sup>

Under normal conditions, the maxillary and ethmoid sinuses are already present, albeit with a reduced volume, at birth.<sup>9</sup> The sphenoid, however, is best identified after 3 or 5 years of age.<sup>10,11</sup> The frontal sinuses, which are the last to form, are absent at birth, and their pneumatization is visible from the 6th year of life onwards.<sup>11</sup> With growth, the paranasal sinuses become progressively pneumatized and take on their definitive appearance by late adolescence.<sup>10,12</sup>

In addition to pneumatization, the incidence of anatomical variants also differs with age. Variants such as Kuhn and sphenothmoidal (Onodi) cells present in early childhood, while the incidence of septal deviation, CB, and Haller cells increases with age.<sup>10</sup>

To date, studies on PCD have focused mainly on the description of the Lund-Mackay (LM) score and the pneumatization of the paranasal sinuses. Those studies have identified a higher prevalence of sinus aplasia and hypoplasia in this population. Due to the progressive pneumatization of the paranasal sinuses, which is completed only in early adulthood, and to the fact that anatomical variability increases with age, we believe a detailed description of paranasal sinus CT findings exclusively in the adult population with PCD would be relevant. On the other hand, in addition to the changes in mucociliary clearance observed in these patients, it is also important to identify sinonasal lesions, and anatomical variants, all of which can contribute to altered drainage of the paranasal sinuses and subsequent development of sinonasal symptoms. The present study is relevant for safe surgical planning, should help maintain heightened diagnostic suspicion of PCD, and proposes a standard to describe CT findings in these patients. To date, we have not identified another study with such detailed tomographic description of adults with PCD.

## Materials and Methods

We conducted a retrospective study of paranasal sinus CT scans of adult (age > 18 years) patients with PCD requested for investigation of CRS at a tertiary hospital, using a non-probabilistic convenience sampling strategy.

According to the European Respiratory Society (ERS) Task Force Guidelines for diagnosing PCD, patients have a confirmed diagnosis of PCD if they present a typical history of PCD and hallmark ciliary ultrastructure defects for PCD assessed by transmission electron microscopy (TEM), or non-ambiguous biallelic mutations in PCD causing genes.<sup>13</sup> The American Thoracic Society guideline considers the diagnosis of PCD to be positive if the patient presents the alterations adopted by the ERS guideline or repeated low nasal nitric oxide (nNO) levels associated with exclusion of the diagnosis of cystic fibrosis.<sup>14</sup> Thus, PCD diagnosis in this sample was based on presence of typical clinical history and of different combinations of the following tests: low nNO measurement or high-speed video analysis showing altered cilia beat frequency and cilia beat pattern, plus homozygosity for a proven PCD variant gene or TEM showing altered cilia ultrastructure consistent with PCD. Finally, some patients also underwent immunofluorescence, demonstrating absence of staining of ciliary proteins.

The present study was approved by the local research ethics committee (approval number: 4.408.630).

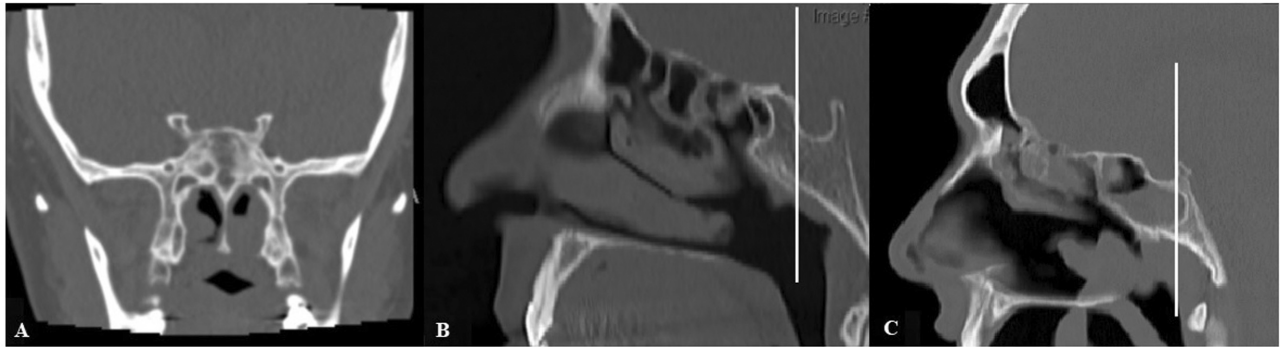
The scans were analyzed by two specialist rhinologists and by a radiologist specializing in head and neck CT.

Computed tomography scans were described according to the characteristics seen in each of the paranasal sinuses. To describe the pneumatization of each sinus, we adopted the classification suggested by Eggesbo et al.<sup>15</sup> for a study conducted in patients with cystic fibrosis (CF). We adopted this classification because it is based on anatomical parameters, which allows its application for safer surgical planning and permits comparison with relevant previous studies.

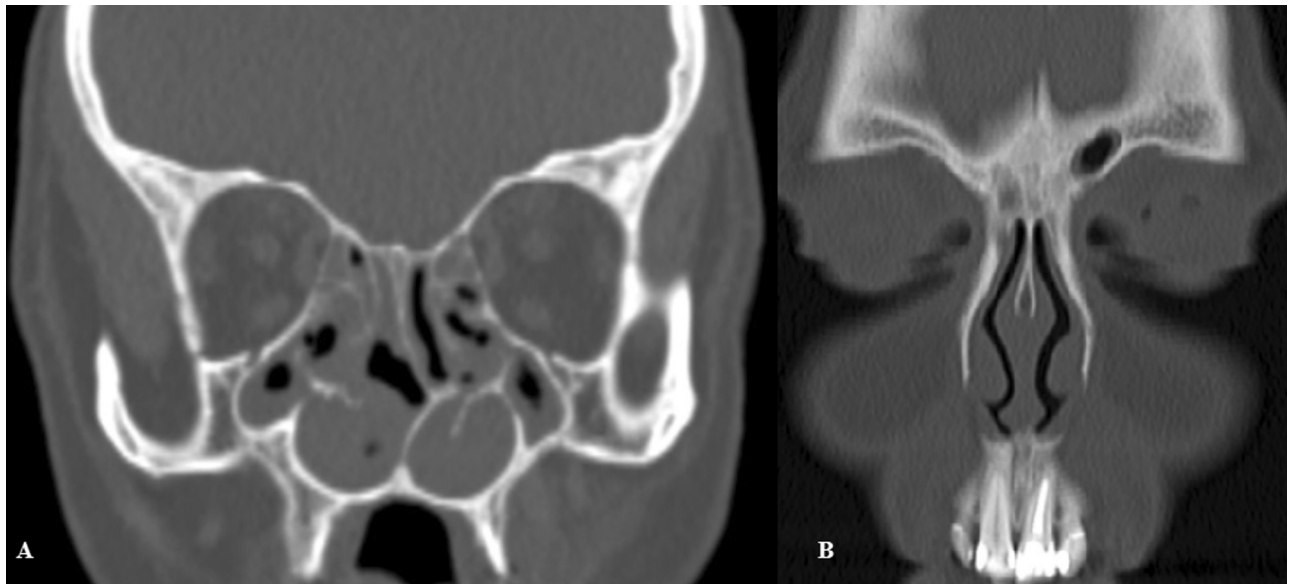
We classified the sphenoid sinus as aplastic when there was no pneumatization posterior to the sphenoidal rostrum,<sup>10</sup> hypoplastic when it was very small and located only in the anterior part of the sphenoid, or fully developed when its pneumatization extended posteriorly beyond a vertical line in the sagittal plane that touches the anterior wall of the pituitary fossa<sup>15</sup> (– Fig. 1).

The frontal sinus was classified as aplastic when it exhibited no pneumatization superior to the frontal beak or to a horizontal line passing through the superior border of the orbit. We classified it as hypoplastic when the superior border of the sinus was smooth, there was no septation within the sinus, and its pneumatization did not extend beyond a vertical line that crossed the midline of the orbit. In cases in which the sinus was pneumatized beyond this vertical line, we classified it as fully developed<sup>10,15</sup> (– Fig. 2).

Maxillary sinuses were classified as aplastic when there was no pneumatization. Hypoplasia was defined when the sinus met four of the following five criteria: oval shape, absence of pneumatization below the level of the nasal floor, enlarged and oval-shaped orbit, medial sinus wall lateral to a



**Fig. 1** (A) Aplasia of sphenoid sinuses. (B) Hypoplasia of sphenoid sinus. (C) Fully developed sphenoid sinus.



**Fig. 2** (A) Hypoplastic maxillary sinuses. (B) Right frontal sinus aplasia and left frontal sinus hypoplasia.

vertical line drawn tangent to the medial border of the orbit (the insertion of the inferior turbinate at the lateral wall of the nose was used as a landmark when sinus surgery had been performed prior to examination), and lateral extension of the sinus medial to a vertical line drawn through the midline of the orbit at the level of the infundibulum, in the coronal plane. Finally, we classified it as fully developed when it did not meet the criteria for hypoplasia.<sup>15</sup>

We also described changes suggestive of osteoma, mucocoele, bone erosion, fungus ball, polyp, and anatomical variations such as concha bullosa (CB), paradoxical middle turbinate (PMT), agger nasi cells, Haller cells, frontoethmoidal cells, and Onodi cells.

We performed Lund-Mackay (LM) classification of all CT scans.<sup>16</sup> As recommended by Lund et al.,<sup>17</sup> if the patient had undergone sinonasal surgery prior to the CT scan, we still classified the sinuses according to their degree of opacification. Partially opacified sinuses were assigned a score of 1; fully opacified sinuses, 2; and sinuses free of opacification, 0. The osteomeatal complex was classified as 0 when free of opacification or open due to previous maxillary antrostomy, and as 2 if blocked or stenotic due to previous antrostomy. For patients

who had aplasia of one or more paranasal sinuses, we assigned a score of 0 for each aplastic sinus<sup>17</sup> and used the rule of three to derive what score would be assigned if the patient had all sinuses. To do so, we multiplied 24 (the maximum score for a patient with all sinuses) by the score actually assigned to the patient and divided the product by the maximum possible score for that particular patient.<sup>18</sup>

Septal deviations were described in terms of presence, location, and contact with the turbinates. Finally, we considered the inferior turbinate hypertrophic if it touched the floor of the nasal cavity at any point and the middle turbinate (MT) hypertrophic if it touched the inferior turbinate.

To identify the severity of lower-airway disease, we considered patients who had a forced expiratory volume in the first second (FEV<sub>1</sub>) of less than 50% of the predicted on diagnostic spirometry as having severe lung disease.<sup>19</sup>

## Results

Twenty-one adult patients with confirmed PCD (13 men and 8 women), aged 18 to 62 years (mean, 38.1 years), were included in the study. Of these, 4 (19%) had undergone endoscopic

**Table 1** Clinical and demographic data from primary ciliary dyskinesia patients

Patient	Age(y)	Sex	Positive tests	FEV <sub>1</sub> < 50%	ESS before CT
1	62	M	Low nNO;HSVA (altered cilia movement); TEM (ODA); PCD gene: DNAI2.	No	Yes
2	37	M	Low nNO; HSVA (altered cilia movement); TEM (MTD + IDA); PCD gene: CCDC40.	No	No
3	34	M	Low nNO; HSVA (altered cilia movement); TEM (MTD + IDA); PCD gene: CCDC40.	No	Yes
4	55	F	Low nNO; HSVA (Altered Cilia Movement); TEM (OIDA).	Yes	No
5	37	F	Low nNO; HSVA (altered cilia movement); TEM (MTD + IDA); PCD gene: CCDC39; IF (CCDC39).	Yes	No
6	18	M	Low nNO; HSVA (altered cilia movement); TEM (MTD + IDA); PCD gene: CCDC40; IF (CCDC39).	Yes	No
7	54	M	Low nNO; HSVA (altered cilia movement); TEM (MTD + IDA); IF (CCDC39).	Yes	Yes
8	30	F	Low nNO; PCD gene: DNAAF3.	No	TIB
9	51	F	Low nNO; PCD gene:RSPH1.	No	No
10	40	M	Low nNO; HSVA (altered cilia movement); TEM (OIDA).	No	No
11	26	M	Low nNO; PCD gene: DNAH11.	Yes	No
12	23	F	Low nNO; HSVA (altered cilia movement); TEM (ODA); PCD gene: DNAH5; IF (DNAH5).	No	No
13	41	M	Low nNO; TEM (OIDA); PCD gene: DNAAF3.	Yes	No
14	18	F	Low nNO; HSVA (altered cilia movement); PCD gene: RSPH1.	No	No
15	54	M	Low nNO; HSVA (altered cilia movement); TEM (ODA); PCD gene: DNAH5.	Yes	No
16	47	M	TEM (ODA); PCD gene: DNAH5.	Yes	No
17	29	F	Low nNO; TEM (MTD + IDA); PCD gene: CCDC40; IF (CCDC39).	Yes	No
18	18	M	Low nNO; TEM (ODA); IF (DNAH5).	No	No
19	37	M	Low nNO; HSVA (altered cilia movement); TEM (MTD + IDA).	Yes	No
20	40	M	HSVA (altered cilia movement); PCD gene: DYX1C1-CCGP1	No	No
21	50	F	Low nNO; HSVA (altered cilia movement); TEM (ACP).	Yes	Yes

Abbreviations: ACP, absence of central pair; ACP + T: absence of central pair and transposition; altered cilia movement, altered cilia beat frequency and cilia beat pattern; CT, computed tomography; ESS, endoscopic sinus surgery; F, female; FEV<sub>1</sub>, forced expiratory volume in the first second of the forced vital capacity; HSVA, high-speed video analysis; IF, cilia immunofluorescence; IT, inferior turbinectomy; M, male; MTD + IDA, microtubular disarrangement and inner dynein arm; nNO, nasal nitric oxide; ODA, outer dynein arm; OIDA, outer inner dynein arm; PCD gene, proved primary ciliary dyskinesia variant gene (homozygotes); TEM, transmission electron microscopy.

endonasal surgery (EES) prior to the CT scan we evaluated (► **Table 1**). Overall, 10 (47.6%) of the patients had situs inversus (SI), 5 (23.8%) were lung transplant recipients, and 1 (4.8%) had undergone lung lobectomy. Severe pulmonary disease was present in 11 (52.4%) patients at the time of diagnosis.

The incidence of aplasia and hypoplasia of each paranasal sinus can be seen in ► **Table 2**.

**Frontal sinus:** Analysis by patient revealed that 11 (52.4%) had frontal aplasia, 5 (23.8%) bilateral and 6 (28.6%) unilat-

eral, all with a hypoplastic contralateral sinus. We identified bilateral hypoplasia in 7 patients (33.3%) and bilateral fully developed in 3 (14.3%).

**Sphenoid sinus:** On the analysis by patient, 5 (23.8%) presented with bilateral fully developed sphenoids and 3 (14.3%) with unilateral fully developed. Aplasia was bilateral in 2 (9.5%) and unilateral in 2 (9.5%). We found bilateral hypoplasia in 9 patients (42.8%) and unilateral hypoplasia in 5 (23.8%).

**Maxillary sinus:** On the analysis by patient, 11 (52.4%) presented with bilateral fully developed maxillary sinuses and 3 (14.3%) with unilateral. Finally, 7 (33.3%) had bilateral hypoplasia.

**Ethmoid sinus:** We observed loss of ethmoid trabeculae in 13 patients (61.9%).

**Lund-Mackay score:** The LM score ranged between 8.7 and 18 (mean, 13.5 and median (interquartile range [IQR]), 14.2 [3.3]). Of the 147 pneumatized sinuses, only 6 (4.1%) had a

**Table 2** Paranasal sinus pneumatization

	Frontal N (%)	Sphenoid N (%)	Maxillary N (%)
<b>Aplasia</b>	16 (38.1)	6 (14.3)	0
<b>Hypoplasia</b>	20 (47.6)	23 (54.8)	17 (40.5)
<b>Fully developed</b>	6 (14.3)	13 (30.9)	25 (59.5)

**Table 3** Lund-Mackay score for each pneumatized sinus, anatomical variations, and lesions

	Maxillary	Ethmoid	Frontal	Sphenoid
	N (%)	N (%)	N (%)	N (%)
LM 0	0 (0)	0 (0)	3 (12)	3 (8.8)
LM 1	37 (88.1)	37 (88.1)	17 (68)	30 (88.2)
LM 2	5 (11.9)	5 (11.9)	5 (20)	1 (2.9)
<b>Anatomical variations n (%)</b>				
<b>Turbinate variants:</b>				
Hypertrophic inferior turbinate: 10 (47.6%)				
Hypertrophic middle turbinate: 8 (38.1%)				
Concha bullosa: 2 (9.5%)				
Paradoxical middle turbinate: 2 (9.5%)				
<b>Agger nasi: 17 (81%)</b>				
<b>Onodi cell: 5 (23.8%)</b>				
<b>Frontoethmoidal cell: 1 (4.8%)</b>				
<b>Marked septal deviation: 10 (47.6%)</b>				
<b>Lesions</b>				
Probable fungus ball: 1 (4.8%)				
Osteoma: 1 (4.8%)				
Mucocele: 1 (4.8%)				
Probable antrochoanal polyp: 1 (4.8%)				
Probable bone erosion: 1 (4.8%)				

Abbreviation: LM, Lund-Mackay.

score of 0; of these, 3 were frontal sinuses and 3 were sphenoid sinuses (►Table 3).

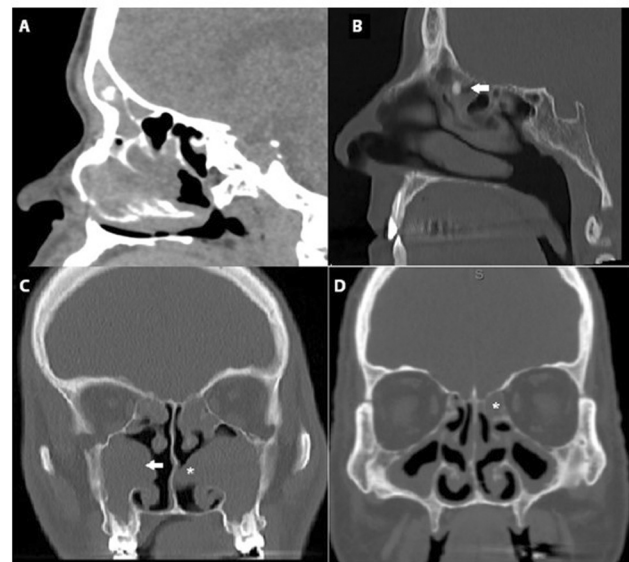
**Gender differences:** Bilateral sphenoid sinus aplasia was identified in 2 men and no women. Likewise, bilateral frontal aplasia was identified in 5 men and in none of the women. However, women and men accounted for 3 cases of unilateral frontal aplasia each. Regarding the maxillary sinus, neither sex had sinus aplasia, but 4 men and 3 women had bilateral hypoplasia. The men's LM score ranged between 9.3 and 17 (mean: 13.7). In women, the score ranged between 8.7 and 18 (mean: 13.1).

**Turbinate variants:** Among the anatomical variations identified in the nasal turbinates, 2 patients (9.5%) had a CB. The inferior turbinate exhibited marked hypertrophy bilaterally in 10 patients (47.6%). On the other hand, 8 (38.1%) patients had marked bilateral hypertrophy of the MT. We found paradoxical curvature of the MT in 2 patients (9.5%).

**Anatomical variations:** We identified agger nasi in 17 patients (81%), Onodi cell in 5 (23.8%), frontoethmoidal cell in 1 (4.8%), and 10 patients (47.6%) had marked septal deviation.

**Lesions:** One patient had a probable frontal fungus ball, one patient had an image suggestive of anterior ethmoid osteoma, and one patient a small mucocele in the left anterior ethmoid. (►Fig. 3).

In one patient who had undergone maxillary antrostomy before CT, we observed a lesion suggestive of an antrochoanal polyp, and the patient was referred for further investigation (►Fig. 3). The same patient had a homogeneous soft-tissue density with rounded borders within the right maxillary sinus, suggesting a cyst or polyp.



**Fig. 3** Images suggestive of: (A) Left frontal sinus fungal ball. (B) Left ethmoid sinus osteoma (arrow). (C) Left maxillary sinus antrochoanal polyp (\*) and a right maxillary sinus cyst (arrow). (D) Left ethmoid sinus mucocele (\*).

**Bone erosion:** We identified an image suggestive of bone erosions in the frontal sinuses of 1 patient (4.8%). These defects were found in the posterior table and floor of the right frontal sinus, and an additional probable erosion was seen in the posterior table of the left frontal sinus (►Fig. 4). This patient underwent EES for treatment of CRS and did not present any evidence of cerebrospinal fluid (CSF) leak.





**Fig. 4** Images suggestive of: (A) Left frontal sinus with posterior table erosion (arrowhead) and right frontal sinus with floor erosion (arrow). (B) Right frontal sinus with floor erosion (arrow). (C) Left frontal sinus with posterior table erosion (arrow).

## Discussion

In the present study, we found a high prevalence of aplastic (38.1%) and hypoplastic (47.6%) frontal sinuses. Overall, 52.4% of patients had frontal sinus aplasia (bilateral in 23.8%). Bilateral hypoplasia was identified in 33.3% of patients. The prevalence of sinus aplasia was higher than that reported by Marino et al., who found that 18.4% of the frontal sinuses of patients without sinonasal disease were entirely absent or did not extend beyond the superior border of the orbit (which, in the present study, was classified as aplasia).<sup>20</sup> Park et al. identified frontal aplasia in 5.3% of volunteers without sinonasal disease, but the authors' definition of aplasia is unclear.<sup>21</sup> Pifferi et al. and Bequignon et al. used the criteria proposed by Orlandi et al.<sup>18</sup> Thus, they defined aplasia as absence of pneumatization of the frontal sinus and hypoplasia if the sinus met the same criteria adopted in the present study. In a sample of children and adults with PCD, Pifferi et al. observed aplasia in 32% of frontal sinuses and hypoplasia in 21%, which was statistically superior to that found in patients with secondary ciliary dyskinesia (SCD) (11% and 17%, respectively). The authors believe that the fact that patients with SCD also have a higher incidence of sinus hypoplasia and aplasia than the normal population could support the hypothesis of an influence of the inflammatory process on sinus underdevelopment.<sup>22</sup> Bequignon et al. evaluated frontal sinus pneumatization in adults with PCD and identified a lower prevalence, with complete aplasia in 17.1% and hypoplasia in 14.6% of patients.<sup>23</sup> El-Sayed et al. identified a 56.3% combined rate of frontal sinus hypoplasia and aplasia in patients with PCD aged 2 years and older. However, the authors did not report clear criteria for aplasia and hypoplasia.<sup>24</sup> Alanin et al., during the preoperative evaluation of adults and children with PCD, identified frontal or sphenoid hypoplasia in 58% of patients, but again did not report clear criteria.<sup>25</sup>

Of the total number of sphenoid sinuses evaluated, we observed aplasia in 14.3% and hypoplasia in 54.8%. Considering the analysis performed per patient, 19% had at least one aplastic sinus. We identified bilateral hypoplasia in 42.8% of patients and unilateral hypoplasia in 23.8%. These percen-

tages are in stark contrast to those described in adults with PCD by Bequignon et al., who found hypoplasia in only 24.4% of patients, and no cases of sphenoid aplasia.<sup>23</sup> In a study of adults and children with PCD, Pifferi et al. found aplastic sphenoid sinuses in 15% and hypoplastic sinuses in 32%. Among patients with SCD, 7% had aplastic sphenoids and 3% had hypoplasia.<sup>22</sup> Both studies used criteria similar to those adopted in the present investigation to classify sphenoid pneumatization.

We found that 40.5% of maxillary sinuses were hypoplastic. Analysis by patients showed bilateral maxillary hypoplasia in 33.3% and unilateral hypoplasia in 14.3%. Pifferi et al. identified hypoplasia in 12% of maxillary sinuses of adults and children with PCD.<sup>22</sup> Pappa et al. identified that 88% of maxillary sinuses of adults with PCD had a smaller volume than the average volume of patients in the control group.<sup>26</sup> Bequignon et al. identified maxillary sinus aplasia in 2.4% of adult patients with PCD and hypoplasia in 4.9%.<sup>23</sup> On the other hand, Lorkiewicz-Muszyńska et al. evaluated the CT scans of 170 children without sinonasal diseases, and all patients had maxillary sinuses.<sup>12</sup> Bequignon et al. and Pifferi et al. adopted criteria similar to those used in the present study and that, if adopted, would not change our findings.

Several factors can be considered to at least partly explain the high prevalence of altered pneumatization of the paranasal sinuses in our sample: our patients were followed up at a tertiary hospital, 28.6% underwent lung surgery, more than half had severe lung disease, and we did not perform CT in patients without sinonasal complaints.

Kayabasi et al. found that patients with frontal, sphenoid, or maxillary sinus hypoplasia have a deeper olfactory fossa and a longer lateral lamella of the cribriform plate. These patients are thus at greater risk of skull base injury during EES.<sup>27</sup> Due to the higher incidence of paranasal sinus hypoplasia and aplasia in adults with PCD, these patients may be at increased risk of iatrogenic skull base injury.

In the present study, the LM score ranged between 8.7 and 18 (mean = 13.5, and median [IQR] = 14.2[3.3]). Pappa et al. identified LM scores between 6 and 16 (mean, 10.6) in adults with PCD, while the mean score in the control group was only 0.7.<sup>26</sup> Frija-Masson et al. found LM with a median of 15 in 50

adults with PCD<sup>6</sup> and Bhatt et al. found median  $\pm$  standard deviation (SD) of  $15.8 \pm 2.6$  in children with PCD.<sup>28</sup> Pifferi et al. identified a median (IQR) score of 9.0 (6.5) in adult and pediatric patients with PCD and 3.5 (7) in patients with SCD, with a statistically significant difference between these groups.<sup>22</sup> Bequignon et al. found that 12% of CT scans were normal, and the mean LM score was  $6.2 \pm 3.6$  in adults with PCD.<sup>4</sup>

In a study of 323 CT scans of patients without sinonasal diseases, Marino et al. identified CB in 28.9% of MTs.<sup>20</sup> Sedaghat et al. identified CB in 41.7% of patients with allergic rhinitis who progressed to CRS.<sup>29</sup> In the present study, 9.5% of the patients had CB and 9.5% had paradoxical curvature of the MT, which can lead to obstruction of the middle meatus. Just as the paranasal sinuses of these patients are less pneumatized than those of individuals without disease, so too may their MTs. Agger nasi cells were identified in 81% of patients, Onodi cells in 23.8%, and frontoethmoidal cells in 4.8%. According to Lund et al., depending on the method of analysis, the incidence of agger nasi cell ranges from 70 to 90% in the literature.<sup>30</sup> These anatomical variations, associated with altered mucociliary clearance, can hinder drainage of the paranasal sinuses. The finding of bilateral inferior turbinate hypertrophy in 47.6% of patients and MT hypertrophy in 38.1% can also contribute to sinonasal obstructive symptoms. Assessing nasal endoscopy in 64 adults with PCD, Bequignon et al.,<sup>4</sup> identified inferior turbinate hypertrophy in only 34.4%.

In the present study, we found that only men had bilateral sphenoid and frontal aplasia, even with a similar mean LM score between genders. Previous studies did not differentiate between genders, but we believe that a study with a larger number of patients can help elucidate whether a real gender difference exists.

Among other lesions, we identified images suggestive of fungus ball within the frontal sinus, antrochoanal polyp, ethmoidal mucocele and ethmoidal osteoma. The frontal sinus is the rarest location for a fungus ball among the paranasal sinuses. In a review of the literature published between 1973 and 2008, Popko et al. identified only 20 cases of frontal sinus fungus ball.<sup>31</sup> Bequignon et al. did not identify lesions such as mucocele on CT scans of 41 adults with PCD.<sup>23</sup> Berlucchi et al. described what they considered to be the first report of ethmoidal mucocele in PCD, and the authors suggested that, in children, the presence of mucoceles should raise diagnostic suspicion of PCD.<sup>32</sup> In a survey of patients who underwent surgery for mucocele over a 10-year period, only 6.5% had exclusively ethmoidal mucocele.<sup>33</sup> Finally, we identified bone dehiscence in the frontal sinuses in 4.8% of patients.

The limitations of this investigation include the lack of a control group for comparison and the small sample size. However, given that PCD is a rare disease and a particularly difficult diagnosis to confirm, it is relevant as one of the largest series in the literature limited to adults with known PCD.

## Conclusion

The present study proposes a standardized description of CT findings in patients with PCD. We identified a high incidence of paranasal sinus hypoplasia and aplasia in adults. Notably, we did not identify a single CT scan in which all paranasal sinuses were completely free. These findings, combined with characteristic clinical picture or even specific questionnaires such as PICADAR,<sup>34</sup> may help heighten diagnostic suspicion of PCD, especially in countries with limited access to diagnostic methods. The presence of anatomical variants, hypertrophy of the middle and lower turbinates, septal deviation and lesions can also contribute to sinonasal symptoms and impair drainage of the paranasal sinuses. The present study highlights the need to consider these findings in the preoperative planning of patients who require surgery.

### Authors' Contribution

All authors contributed to the study conception and design. Material preparation and data collection were performed by Diogo Plantier. Computed tomography scans analyses were performed by Diogo Plantier, Renata Pilan, and Eloisa Gebrim. The first draft of the manuscript was written by Diogo Plantier, and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript.

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### Conflict of Interests

The authors declare that there are no competing financial or nonfinancial interest and there are no personal relationships with other people or organizations that could inappropriately influence (bias) their work.

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