RESEARCH ARTICLE
Follow-up practices in Italian primary ciliary dyskinesia centers: a national survey

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ABSTRACT

Primary ciliary dyskinesia (PCD) is a rare, chronic, multisystem disorder requiring lifelong respiratory and multidisciplinary care. A previous Italian survey described diagnostic and therapeutic practices, whereas information on longitudinal follow-up remains limited. We performed a descriptive secondary analysis of a national survey conducted between October 2020 and June 2021 among Italian PCD centers, focusing on respiratory and multidisciplinary follow-up. Centers were classified by patient age group and geographical location. Twenty of 22 centers responded, representing 12 of 20 Italian regions; 4 were pediatric-only, 15 mixed pediatric-adult, and 1 adult-only. Clinical visits and pulmonary function tests were performed at least three times per year in 70% of centers, while respiratory physiotherapy was reassessed with the same frequency in 55% of centers. Chest computed tomography was most commonly performed every 3-4 years (60%). Sputum culture was obtained at each visit in 50% of centers. Multidisciplinary follow-up was variable: audiometry was performed according to clinical status in 50% of centers; fertility assessment was reported by 60%, cardiological assessment by 95%, and psychological support by 75%. Most comparisons by center type and geographical area did not reach statistical significance; however, exploratory analyses identified differences in sputum culture practices according to center type ($p = 0.003$).

Clinical visits and pulmonary function testing were frequently reported, while variability was observed in microbiological surveillance, imaging, physiotherapy reassessment, and multidisciplinary care. These findings support the development of shared national minimum

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standards for longitudinal PCD care while preserving individualized, age-appropriate management.

IMPACT STATEMENT

This national survey highlights variability in longitudinal PCD care across Italian centers and supports shared minimum standards for multidisciplinary, age-appropriate follow-up.

HIGHLIGHTS BOX

What is already known about this topic? Primary ciliary dyskinesia requires lifelong respiratory and multidisciplinary care, but the organization and frequency of longitudinal follow-up remain incompletely standardized across centers. **What does this article add to our knowledge?** This national survey shows relatively consistent clinical and lung function monitoring across Italian PCD centers, but greater variability in microbiological surveillance, imaging, physiotherapy reassessment, and multidisciplinary care. **How does this study impact current management guidelines?** These findings suggest the need for shared national minimum standards for PCD follow-up, while allowing care to remain individualized according to age, clinical status and patient needs.

KEY WORDS

Follow-up; multidisciplinary care; primary ciliary dyskinesia; rare disease; respiratory monitoring.

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10.56164/PediatrRespirJ.2026.21

INTRODUCTION

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Primary ciliary dyskinesia (PCD; MIM 244400) is a rare, chronic and genetically heterogeneous disorder of motile cilia, characterized by impaired mucociliary clearance and recurrent, often chronic, respiratory tract disease (1). Its clinical spectrum extends beyond the lower airways and may include chronic rhinosinusitis, otologic disease, bronchiectasis, laterality defects and fertility-related issues (2). For this reason, PCD requires long-term, multidisciplinary care aimed at monitoring respiratory morbidity, preserving lung function, detecting infectious complications and addressing extrapulmonary manifestations. Recent work has specifically addressed follow-up in children with PCD, further highlighting the need for structured, age-appropriate longitudinal care (3). Early diagnosis remains a key component of PCD management (4). However, diagnosis is often delayed, and the organization of care may vary substantially among centers, particularly in healthcare systems where PCD management is not fully centralized. Differences in local expertise, available diagnostic tools, patient volume and access to multidisciplinary services may influence both the timing and content of follow-up assessments. A previous Italian multicenter survey provided updated information on the distribution of PCD centers in Italy and described variability in diagnostic procedures and treatment strategies (5). In that survey, 20 of 22 contacted centers responded, representing 12 of the 20 Italian regions, and most centers followed both pediatric and adult patients. Although diagnostic pathways have been progressively standardized through European Respiratory Society recommendations (6), and randomized evidence has begun to emerge for selected treatments, including maintenance azithromycin (7), the organization and timing of longitudinal follow-up remain less uniformly defined. The present study aims to describe respiratory and multidisciplinary follow-up practices in Italian PCD centers, including the frequency of clinical visits, pulmonary function testing, respiratory physiotherapy reassessment, microbiological surveillance, chest imaging, and audiological monitoring, as well as the reported provision of fertility and cardiological

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assessments and availability of psychological support. Furthermore, we explore differences according to center type (pediatric-only, mixed pediatric-adult, and adult-only) and geographical area (Northern, Central, and Southern Italy). By focusing specifically on longitudinal follow-up, this secondary analysis provides additional findings beyond the diagnostic and therapeutic aspects addressed in the previous publication (5).

MATERIALS AND METHODS

Study design and population

Italian patients with PCD are usually followed in pediatric pulmonology centers. A group of PCD experts, in agreement with the Italian patient association Associazione Italiana Discinesia Ciliare Primaria Sindrome di Kartagener (A.I.D. Kartagener Onlus), designed a survey questionnaire to be sent to all Italian pediatric PCD centers. To involve the largest possible number of care centers, members of the Italian Pediatric Respiratory Society (SIMRI) and all acknowledged regional referral centers for PCD and cystic fibrosis were contacted. This study is a descriptive secondary analysis of data collected through a national survey of Italian PCD centers between October 2020 and June 2021. The original survey was previously used to describe the distribution of PCD centers and diagnostic and therapeutic practices in Italy (5). The present analysis specifically addresses respiratory and multidisciplinary follow-up variables, which were not the main focus of the previous report.

Questionnaire

The questionnaire was in Italian and included items on patients' characteristics, diagnostic procedures, treatment strategies, and follow-up practices. Follow-up items included data on

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scheduled clinical visits, pulmonary function testing, respiratory physiotherapy, microbiological surveillance, chest imaging, audiological assessment, fertility evaluation, cardiological evaluation, and psychological support. Audiometry was assessed in terms of examination frequency, whereas fertility and cardiological assessments were recorded as yes/no responses without information on their periodicity. Psychological support was assessed in terms of service availability rather than scheduled evaluations.

The original questionnaire is provided as **Supplementary material**.

Center classification and geographic grouping

Centers were classified according to the age group of patients followed: pediatric centers, mixed pediatric-adult centers, and adult centers. For geographic analyses, centers were grouped into Northern, Central, and Southern Italian macroareas according to their geographical location. The geographic analysis included 10 Northern centers, 4 Central centers, and 6 Southern centers.

Statistical analysis

Results are presented as absolute numbers and percentages. Comparisons between center types and geographical macroareas were performed using Fisher-Freeman-Halton exact tests. Given the small sample size and especially the presence of only one adult-only center, all comparisons were considered exploratory. The survey collected center-level information on clinical practice and did not include identifiable or individual-level patient data. Therefore, formal Ethics Committee approval and individual informed consent were not required according to the applicable institutional requirements.

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RESULTS

Participating centers and study population

Twenty of 22 contacted centers responded to the survey (91%), representing 12 of the 20 Italian regions. PCD care was not centralized within the Italian National Health Service during the study period, and participating centers were unevenly distributed across the country. Four centers (20%) were pediatric-only, 15 (75%) followed both pediatric and adult patients, and one (5%) was adult-only. Fourteen centers (70%) were also involved in the management of patients with cystic fibrosis.

The number of patients followed and the reported mean age at diagnosis varied among centers, as summarized in **Table 1**.

The geographical distribution of participating centers according to center type is shown in **Figure 1**.

Distribution of pediatric and adult patients in mixed centers

Among the 15 mixed pediatric-adult centers, the proportion of pediatric patients varied widely, ranging from 13% at the University Hospital of Verona to 90% at De Marchi Hospital, the University Hospital of Padua, and the University of Catania. Conversely, the adult component ranged from 10% to 87%. Some mixed centers were predominantly pediatric, whereas others followed a mainly adult population despite being classified as mixed centers. A balanced distribution was observed at Ca' Granda Ospedale Maggiore, where children and adults each accounted for 50% of followed patients. The complete center-level distribution is shown in **Figure 2**.

Diagnostic and treatment overview

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Diagnostic procedures and treatment strategies among participating centers have been described in detail in our previous publication (5). Importantly, reported treatment use refers to the proportion of centers prescribing a given treatment to at least some patients, rather than to routine administration to all patients. These findings are summarized in **Table 2** to provide clinical context for the present analysis, which focuses specifically on respiratory and multidisciplinary follow-up practices.

Respiratory follow-up

Respiratory follow-up practices across all participating centers are summarized in **Table 3**. Clinical visits and pulmonary function tests were performed at least three times per year in 14/20 centers (70%). Five centers (25%) performed these assessments twice yearly, while 1/20 center (5%) reported annual assessment. Respiratory physiotherapy was reassessed at least three times per year in 11/20 centers (55%), twice yearly in 8/20 (40%), and once yearly in 1/20 (5%). Greater variability was observed in chest imaging practices. Chest X-ray was most frequently performed every 3-4 years (7/20 centers, 35%), followed by a single assessment (5/20, 25%), annual imaging (4/20, 20%), assessment according to clinical status (3/20, 15%), and every 2 years (1/20, 5%). Chest computed tomography (CT) was commonly scheduled every 3-4 years (60%). Microbiological surveillance also showed variable approaches. Sputum culture was performed at each visit in 10/20 centers (50%), according to clinical status in 6/20 (30%), twice yearly in 2/20 (10%), and four times per year in 2/20 (10%). Overall, clinical visits and pulmonary function testing showed the most consistent respiratory follow-up pattern, whereas imaging and microbiological surveillance were characterized by greater variability in timing and frequency.

Patterns according to center type

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Respiratory follow-up according to center type is summarized in **Table 3**.

Clinical visits and pulmonary function tests followed a similar pattern across center types. Among pediatric-only centers, 3/4 (75%) reported at least three assessments per year and 1/4 (25%) one assessment per year. Among mixed centers, 10/15 (66.7%) reported at least three assessments per year and 5/15 (33.3%) two assessments per year. The single adult-only center reported at least three clinical visits and pulmonary function assessments per year. Respiratory physiotherapy reassessment at least three times per year was reported by 2/4 pediatric-only centers (50%), 8/15 mixed pediatric-adult centers (53.3%), and the single adult-only center. Chest imaging practices were more heterogeneous. Pediatric centers reported chest X-ray either once or annually, whereas mixed centers most frequently reported chest X-ray every 3-4 years (7/15, 46.7%). Chest CT was performed every 3-4 years in 10/15 mixed centers (66.7%) and in the adult-only center, whereas 2/4 pediatric centers (50%) performed CT according to clinical status. Marked descriptive differences were also observed in microbiological surveillance. Most comparisons did not reach statistical significance. However, sputum culture practices differed significantly according to center type ($p = 0.003$). All four pediatric-only centers reported obtaining sputum cultures according to clinical status, whereas 10/15 mixed pediatric-adult centers (66.7%) reported obtaining cultures at each visit. The single adult-only center reported performing sputum cultures four times per year. These findings should be interpreted cautiously, given the limited number of participating centers, particularly the presence of only one adult-only center.

Patterns according to geographic area

Respiratory follow-up according to geographical macroarea is summarized in **Table 4**. Clinical visits and pulmonary function testing were performed at least three times per year by all Southern centers (6/6, 100%), compared with 6/10 Northern centers (60%) and 2/4 Central

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centers (50%). Respiratory physiotherapy reassessment showed geographical variability, with assessments at least three times per year reported by 5/6 Southern centers (83.3%), 3/4 Central centers (75%), and 3/10 Northern centers (30%). Chest X-ray schedules varied across geographical areas. Annual chest X-ray was reported mainly by Southern centers (3/6, 50%), whereas Northern centers more frequently reported chest X-ray every 3–4 years (4/10, 40%), once only (3/10, 30%), or according to clinical status (2/10, 20%). Chest CT was performed every 3-4 years in 5/6 Southern centers (83.3%), 2/4 Central centers (50%), and 5/10 Northern centers (50%). Other CT schedules were reported by 2/10 Northern centers (20%) and 1/6 Southern center (16.7%). Microbiological surveillance also showed descriptive differences across geographical areas. Sputum culture at each visit was reported by 3/4 Central centers (75%), 3/6 Southern centers (50%), and 4/10 Northern centers (40%). Culture according to clinical status was reported by 3/10 Northern centers (30%), 1/4 Central centers (25%), and 2/6 Southern centers (33.3%). Twice-yearly cultures were reported only by Northern centers (2/10, 20%), while four cultures per year were reported by one Northern center (10%) and one Southern center (16.7%). None of the comparisons reached statistical significance.

Multidisciplinary follow-up

Multidisciplinary follow-up practices across participating centers are summarized in **Table 5**. Audiometry was performed according to clinical status in 10/20 centers (50%), every 1-2 years in 9/20 (45%), and only once in 1/20 (5%). Fertility and cardiological assessments were reported by 12/20 (60%) and 19/20 centers (95%), respectively, while psychological support services were available in 15/20 centers (75%). Unlike audiometry, the questionnaire did not assess the frequency of fertility or cardiological evaluations, and psychological support referred to service availability rather than scheduled assessments.

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Patterns according to center type

Multidisciplinary follow-up according to center type is summarized in **Table 5**.

No statistically significant differences were observed. Audiometry was scheduled every 1-2 years in 3/4 pediatric-only centers (75%) and in the single adult-only center. In contrast, mixed pediatric-adult centers most commonly performed audiometry according to clinical status (9/15, 60%), while 5/15 (33.3%) scheduled assessment every 1-2 years and 1/15 (6.7%) reported a single assessment. Fertility assessment was reported by 2/4 pediatric-only centers (50%) and 10/15 mixed centers (67%), but not by the adult-only center. Cardiological assessment was reported by all pediatric-only centers, 14/15 mixed centers (93%), and the adult-only center. Psychological support was available in 3/4 pediatric-only centers (75%) and 12/15 mixed centers (80%), but was not reported by the adult-only center.

Patterns according to geographic area

Multidisciplinary follow-up according to geographical macroarea is summarized in **Table 6**. No statistically significant differences were detected. Audiometry every 1-2 years was most frequently reported by Central centers (3/4, 75%), compared with Southern (3/6, 50%) and Northern centers (3/10, 30%). In contrast, audiometry according to clinical status was more commonly reported by Northern (6/10, 60%) and Southern centers (3/6, 50%), compared with 1/4 Central center (25%). Fertility assessment was reported by 5/10 Northern centers (50%), 3/4 Central centers (75%), and 4/6 Southern centers (66.7%). Cardiological assessment was almost universal, being reported by 9/10 Northern centers (90%) and by all Central and Southern centers (100%). Psychological support showed greater descriptive variability, being available in 6/10 Northern centers (60%), all Central centers (100%), and 5/6 Southern centers (83%).

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DISCUSSION

This national survey provides a descriptive overview of follow-up practices reported by Italian PCD centers during 2020-2021, complementing our previous analysis of diagnostic and therapeutic approaches (5). The main findings were the frequent use of clinical visits and pulmonary function testing, alongside greater variability in respiratory physiotherapy reassessment, microbiological surveillance, chest imaging, and multidisciplinary care. However, these findings should be interpreted cautiously, and the absence of statistically significant differences in other comparisons should not be considered evidence of equivalent care across centers. Clinical visits and pulmonary function tests were performed at least three times per year in 70% of centers, indicating that regular respiratory monitoring was frequently reported during the study period. Regular respiratory review is central to PCD care, as pulmonary disease may progress even during periods of apparent clinical stability (1-3), and the frequency observed here is broadly consistent with international recommendations. The PCD Foundation consensus recommends regular specialist review and serial lung function assessment (8), while more recent standards of care propose review every three months in children and clinically driven review in adults, with at least annual multidisciplinary assessment (9). The FOLLOW-PCD initiative similarly promotes standardized longitudinal assessments (10). Longitudinal observations by Magnin *et al*. further support repeated functional monitoring, demonstrating progressive deterioration in lung function and structural abnormalities over time (11). Respiratory physiotherapy showed some variability, with reassessment at least three times per year in 55% of centers, twice yearly in 40%, and once yearly in 5%. This should be distinguished from airway clearance itself, which all centers reported as part of daily treatment. Current standards recommend an individualized regimen with formal reassessment at least at the annual review (9). Previous Italian data highlighted organizational differences among respiratory physiotherapy services (12), while

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multidisciplinary management recommendations emphasize the importance of regular, individualized physiotherapy interventions (13). Microbiological surveillance also varied among centers. Although sputum cultures were obtained at each visit in half of the participating centers, other centers followed fixed schedules or clinical indications. Regular microbiological monitoring remains important because respiratory pathogen distribution may change with age and disease progression (14), and international recommendations emphasize standardized microbiological surveillance and infection control (15). Chest imaging practices were similarly heterogeneous, with CT every 3-4 years reported by 60% of centers. Given concerns regarding cumulative radiation exposure, alternative imaging techniques such as magnetic resonance imaging may offer additional monitoring opportunities (16). Sputum culture practices differed significantly according to center type ($p = 0.003$), with pediatric-only centers adopting a clinically driven approach and mixed pediatric-adult centers more frequently performing cultures at each visit. Multidisciplinary follow-up revealed further variability, particularly in audiological monitoring, fertility assessment, and psychological support. This is relevant because PCD is a multisystem disorder requiring attention to extrapulmonary manifestations. Otologic complications are common, particularly during childhood, supporting regular audiological monitoring (17). Longitudinal evidence also suggests age-related changes in hearing impairment, emphasizing the importance of individualized surveillance (18). Fertility assessment was reported by 60% of centers, although reproductive difficulties may affect both male and female patients with PCD (19). Previous research has highlighted gaps in fertility counseling and specialist referrals, supporting greater attention to reproductive health during transition to adult care (20). Cardiological assessment was reported by almost all centers, while psychological support services were available in 75%. Given the potential psychosocial impact of PCD during adolescence (21) and the documented burden of anxiety and depressive symptoms among

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patients and caregivers (22), psychological support represents an important component of multidisciplinary care. However, a fundamental distinction must be made between service availability and regularly scheduled assessments. The questionnaire recorded whether fertility and cardiological evaluations were performed, without investigating their periodicity, while psychological support was assessed only in terms of service availability. Therefore, these findings cannot establish whether multidisciplinary assessments were systematically integrated into follow-up pathways. Several limitations should be acknowledged. First, the survey included only 20 centers, with four pediatric-only centers and a single adult-only center, resulting in limited statistical power and restricting the generalizability of comparisons. Consequently, the absence of significant differences should not be interpreted as evidence that centers provide similar care. Furthermore, multiple comparisons were performed without adjustment, increasing the possibility of chance findings. The results were based on self-reported, center-level information rather than verified clinical records, and therefore may not accurately reflect individual patient management. The absence of patient-level data prevented evaluation of follow-up practices according to age, disease severity, lung function, microbiological status, or clinical outcomes. Differences in resources, staffing, and referral pathways were also not systematically investigated. The survey was conducted between October 2020 and June 2021, during the COVID-19 pandemic, which may have influenced follow-up schedules and the organization of care across participating centers. Notably, 14 of the 20 participating centers (70%) were also involved in cystic fibrosis care. Although experience in managing chronic respiratory diseases may have influenced the organization of PCD follow-up, the survey did not allow us to determine whether follow-up practices differed between centers involved and not involved in cystic fibrosis care. Despite these limitations, this survey identifies areas of variability that may inform the development of shared national minimum standards for PCD follow-up. Such standards should define

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essential components of respiratory and multidisciplinary monitoring while allowing individualized schedules according to patient age, clinical characteristics, and available resources. Future prospective, patient-level studies are needed to assess whether more standardized follow-up pathways improve clinical outcomes and quality of life.

CONCLUSIONS

This study provides an overview of follow-up practices for PCD in Italian referral centers, based on data collected through a national survey conducted between October 2020 and June 2021. Clinical review and pulmonary function testing were frequently reported components of respiratory follow-up, with three or more assessments per year reported by 70% of centers. In contrast, greater variability was observed in respiratory physiotherapy reassessment, microbiological surveillance and chest imaging. Multidisciplinary care was also unevenly structured: cardiological assessment was almost universal, and psychological support was available in most centers, whereas fertility assessment and the timing of audiological surveillance were less consistently implemented. Although most comparisons according to center type and geographical area did not reach statistical significance, sputum culture practices differed significantly according to center type ($p = 0.003$). Nevertheless, the small number of participating centers, particularly the presence of only one adult-only center, precludes definitive conclusions regarding differences in care practices or the superiority of any specific model of care. Overall, these findings highlight differences in follow-up practices among Italian PCD centers and suggest the need for more consistent approaches to patient care. A shared national follow-up pathway could define minimum standards for respiratory review, pulmonary function testing, microbiological sampling, physiotherapy reassessment, imaging and multidisciplinary care, while allowing schedules to be adapted to age, clinical

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status and individual patient needs. Prospective patient-level studies will be needed to determine whether greater standardization of follow-up translates into improved clinical outcomes and quality of life.

COMPLIANCE WITH ETHICAL STANDARDS

Conflicts of interest

The authors declare no conflicts of interest.

Financial support

None.

Author contributions

RC, NU, NI, MA: conceptualization, project administration, methodology, writing – original draft. NI, MA: formal analysis. NU, RC: supervision, writing – review & editing. NU, NI, MA: visualization, writing – review & editing. All authors: data curation, final approval.

Ethical approval

Human studies and subjects

N/A.

Data sharing and data accessibility

The data that support the findings of this review are available from the corresponding author upon reasonable request. Supplementary materials are available at the following link: http://www.pediatric-respiratory-journal.com/wp-content/uploads/2026/11/Supplement_Ullman.pdf.

Publication ethics

Plagiarism

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Authors declare no potentially overlapping publications with the content of this manuscript and all original studies are cited as appropriate.

Data falsification and fabrication

All the data corresponds to the real.

ACKNOWLEDGMENTS

This study was planned in collaboration with the Italian Association A.I.D. Kartagener Onlus and was facilitated by the recent creation of the national research group "PCD and Non-CF Bronchiectasis" within SIMRI. Bambino Gesù Children's Hospital in Rome and Federico II University in Naples are part of the European Reference Network for Rare Lung Disease (ERN-Lung), Primary Ciliary Dyskinesia Network.

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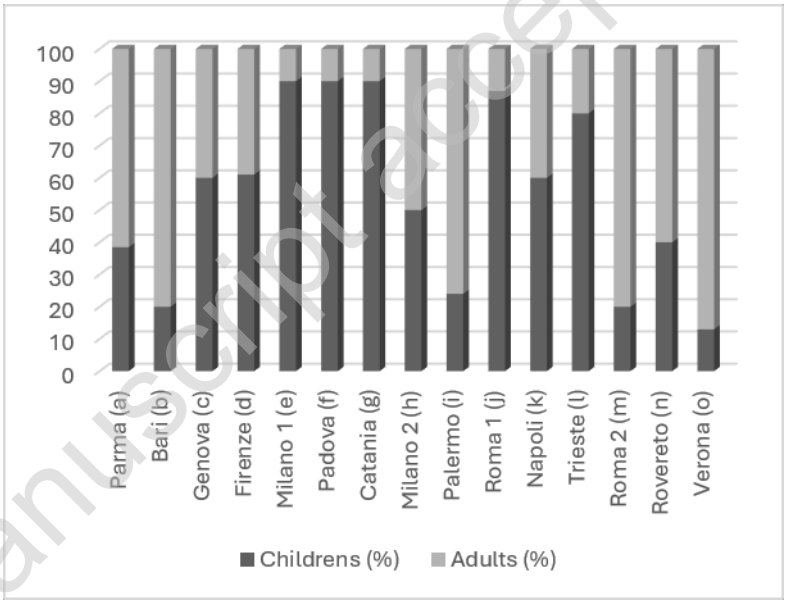
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Figure 1. *Geographical distribution of participating Italian PCD centers.*



PCD: primary ciliary dyskinesia; triangle: pediatric center; square: adult center; circle: mixed pediatric-adult center.

Figure 2. *Distribution of pediatric and adult patients across mixed centers (n = 15).*



(a) Pietro Barilla Children's Hospital, University of Parma; (b) University Hospital of Bari; (c) G. Gaslini Children's Hospital, Genoa; (d) Meyer Children's University Hospital, Florence; (e) De Marchi Hospital, Milan; (f) University Hospital of Padua; (g) University Hospital of Catania; (h) Ca' Granda Ospedale Maggiore, Milan; (i) Children's Hospital G. Di Cristina, Palermo; (j) Bambino Gesù Children's Hospital, Rome; (k) Federico II, University Hospital, Naples; (l) IRCSS Burlo Garofolo, Trieste; (m) Umberto I, Sapienza University of Rome; (n) Hospital of Rovereto; (o) Verona University Hospital.

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Table 1. *Characteristics of participating centers and study population.*

Characteristic	**n (%)**
Participating centers	20 (91)
Italian regions represented	12 (60)
Centers divided by number of patients	
< 10	6 (30)
11-20	5 (25)
21-40	7 (35)
> 40	2 (10)
Centers divided by reported mean age at diagnosis	
< 3 years	3 (15)
4-8 years	9 (45)
9-12 years	5 (25)
13-18 years	2 (10)
> 18 years	1 (5)

Table 2. *Diagnostic procedures and treatment strategies performed in the Italian PCD centers.*

Strategies	**n (%)**
Diagnostic procedure	
Nasal nitric oxide	15 (75)
Nasal brush only	12 (60)
Nasal and bronchial brush	5 (25)
Transmission electron microscopy	18 (90)
High speed video microscopy	8 (40)
Immunofluorescence	5 (25)
Genetic analysis	11 (55)
Treatment	
Airway clearance techniques as daily treatment	20 (100)
Isotonic and/or hypertonic saline solution	20 (100)
Mucolytics (nasal or inhaled)	10 (50)
Inhaled corticosteroids	20 (100)
Prophylactic antimicrobial therapy	19 (95)

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Table 3. *Respiratory follow-up according to type of PCD center.*

Follow-up practice	Frequency / practice	All centers n (%)	Pediatric centers n (%)	Pediatric-adult centers n (%)	Adult center n (%)	P-value
Clinical visits	1/year	1 (5)	1 (25)	0 (0)	0 (0)	0.243
	2/year	5 (25)	0 (0)	5 (33)	0 (0)	
	≥ 3/year	14 (70)	3 (75)	10 (67)	1 (100)	
Pulmonary function tests	1/year	1 (5)	1 (25)	0 (0)	0 (0)	0.243
	2/year	5 (25)	0 (0)	5 (33)	0 (0)	
	≥ 3/year	14 (70)	3 (75)	10 (67)	1 (100)	
Respiratory physiotherapy	1/year	1 (5)	1 (25)	0 (0)	0 (0)	0.367
	2/year	8 (40)	1 (25)	7 (47)	0 (0)	
	≥ 3/year	11 (55)	2 (50)	8 (53)	1 (100)	
Chest X-ray	Once	5 (25)	2 (50)	3 (20)	0 (0)	0.072
	Annually	4 (20)	2 (50)	1 (6.7)	1 (100)	
	Every 2 years	1 (5)	0 (0)	1 (6.7)	0 (0)	
	Every 3-4 years	7 (35)	0 (0)	7 (46.6)	0 (0)	
	According to clinical status	3 (15)	0 (0)	3 (20)	0 (0)	
Chest CT	Once	2 (10)	1 (25)	1 (6.7)	0 (0)	0.172
	Every 3-4 years	12 (60)	1 (25)	10 (66.6)	1 (100)	
	According to clinical status	3 (15)	2 (50)	1 (6.7)	0 (0)	
	Other	3 (15)	0 (0)	3 (20)	0 (0)	
Sputum culture	At each visit	10 (50)	0 (0)	10 (67)	0 (0)	0.003
	2/year	2 (10)	0 (0)	2 (13)	0 (0)	
	4/year	2 (10)	0 (0)	1 (7)	1 (100)	
	According to clinical status	6 (30)	4 (100)	2 (13)	0 (0)	

CT: computed tomography; PCD: primary ciliary dyskinesia. Data are reported as numbers and percentages. The number of centers is: all centers n = 20, pediatric centers n = 4, mixed pediatric-adult centers n = 15, adult center n = 1. P-values between pediatric, mixed and adult centers were calculated using Fisher-Freeman-Halton exact tests.

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Table 4. *Respiratory follow-up practices according to geographic distribution of the centers.*

Follow-up practice	Frequency / practice	Northern Italian centers n (%)	Central Italian centers n (%)	Southern Italian centers n (%)	P-value
Clinical visits	1/year	1 (10)	0 (0)	0 (0)	0.277
	2/year	3 (30)	2 (50)	0 (0)	
	≥3/year	6 (60)	2 (50)	6 (100)	
Pulmonary function tests	1/year	1 (10)	0 (0)	0 (0)	0.277
	2/year	3 (30)	2 (50)	0 (0)	
	≥3/year	6 (60)	2 (50)	6 (100)	
Respiratory physiotherapy	1/year	1 (10)	0 (0)	0 (0)	0.212
	2/year	6 (60)	1 (25)	1 (17)	
	≥ 3/year	3 (30)	3 (75)	5 (83)	
Chest X-ray	Once	3 (30)	2 (50)	0 (0)	0.273
	Annually	1 (10)	0 (0)	3 (50)	
	Every 2 years	0 (0)	0 (0)	1 (16.7)	
	Every 3-4 years	4 (40)	2 (50)	1 (16.7)	
	According to clinical status	2 (20)	0 (0)	1 (16.6)	
Chest CT	Once	1 (10)	1 (25)	0 (0)	0.724
	Every 3-4 years	5 (50)	2 (50)	5 (83)	
	According to clinical status	2 (20)	1 (25)	0 (0)	
	Other	2 (20)	0 (0)	1 (17)	
Sputum culture	At each visit	4 (40)	3 (75)	3 (50)	0.938
	2/year	2 (20)	0 (0)	0 (0)	
	4/year	1 (10)	0 (0)	1 (17)	
	According to clinical status	3 (30)	1 (25)	2 (33)	

CT: Computed Tomography. Data are reported as numbers and percentages. The number of centers is: Northern Italian n = 10, Central Italian n = 4, Southern Italian n = 6. P-values were calculated using Fisher-Freeman-Halton exact tests. Given the small sample size, results should be interpreted as exploratory.

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Table 5. *Multidisciplinary follow-up practices according to center type.*

Follow-up practice	All centers n (%)	Pediatric centers n (%)	Pediatric-adult centers n (%)	Adult center n (%)	P-value
Audiometry					0.380
Once	1 (5)	0 (0)	1 (6.7)	0 (0)	
Every 1-2 years	9 (45)	3 (75)	5 (33.3)	1 (100)	
According to clinical status	10 (50)	1 (25)	9 (60)	0 (0)	
Fertility assessment	12 (60)	2 (50)	10 (67)	0 (0)	0.398
Cardiology assessment	19 (95)	4 (100)	14 (93)	1 (100)	1.000
Psychology service	15 (75)	3 (75)	12 (80)	0 (0)	0.278

Data are reported as numbers and percentages. The number of centers is: all centers n = 20, pediatric centers n = 4, mixed pediatric-adult centers n = 15, adult center n = 1. P-values between pediatric, mixed and adult centers were calculated using Fisher-Freeman-Halton exact tests.

Table 6. *Multidisciplinary follow-up practices according to geographic distribution of the centers.*

Follow-up practice	Northern Italian centers n (%)	Central Italian centers n (%)	Southern Italian centers n (%)	P-value
Audiometry				0.589
Once	1 (10)	0 (0)	0 (0)	
Every 1-2 years	3 (30)	3 (75)	3 (50)	
According to clinical status	6 (60)	1 (25)	3 (50)	
Fertility assessment	5 (50)	3 (75)	4 (67)	0.717
Cardiology assessment	9 (90)	4 (100)	6 (100)	1.000
Psychology service	6 (60)	4 (100)	5 (83)	0.420

Data are reported as numbers and percentages. The number of centers is: Northern Italian n = 10, Central Italian n = 4, Southern Italian n = 6. P-values were calculated using Fisher-Freeman-Halton exact tests.

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