Interstitial lung disease in the intensive unit care setting

Interstitial lung diseases (ILD) are a heterogeneous group of disorders.[1] Although the disease remains stable in some patients, episodes of acute respiratory failure (ARF) requiring invasive mechanical ventilation (IMV) are observed.[2] Acute respiratory failure is often the result of disease progression or an acute exacerbation, but occasionally occurs as an inaugural manifestation or as an adverse reaction to treatment.[1,3]

We conducted a retrospective cohort study including patients admitted into ICU, with previously known ILD diagnosis and as an inaugural event, between January 2004 and May 2015, in order to evaluate the clinical outcome, overall survival and prognostic factors of ILD patients in the ICU setting.

Thirty seven patients were included, 27 (73%) were male. Mean age of 65.1 ± 11.1 years (min: 27; max: 83).

Thirty (81.1%) patients were admitted with previous ILD diagnosis. The diagnoses of the patients admitted in ICU were: 5 (13.5%) idiopathic pulmonary fibrosis (IPF); 5 (13.5%) silicosis; 5 (13.5%) fibrotic unclassifiable ILD; 5 (13.5%) small vessel vasculitis (ANCA +); 4 (10.8%) chronic hypersensitivity pneumonitis; 3 (8.2%) sarcoidosis; 2 (5.5%) rheumatoid lung (RL); 2 (5.5%) acute interstitial pneumonitis; 2 (5.5%) unclear ILD; 2 (5.5%) cryptogenic organizing pneumonia (COP); 1 (2.7%) scleroderma lung and 1 (2.7%) dermatomyositis.

Seventeen had received previous therapy with corticosteroids, 8 immunosuppressant therapy and 10 long term oxygen therapy.

The median length of ICU and hospital stay were 10 days (min: 1; max: 64) and 21 days (min:1; max:100) respectively. Mean APACHE II score was 18.3 ± 7 and mean SAPS II was 36.9 ± 11.5.

Four lung biopsies were performed: 1 surgical lung biopsy and 3 core needle biopsies guided by CT scan. Two of the biopsies were consistent with acute interstitial pneumonitis, 1 with COP and 1 was unclear.

Seventeen (45.9%) patients had acute exacerbation and twenty experienced ARF due to rapid deterioration of disease associated with respiratory infection.

Thirty three patients (89.2%) required IMV, with an median duration of 10 days (min: 1; max: 62). Nine (24.3%) underwent non-invasive ventilation prior to IMV, 6 required tracheostomy and 3 extracorporeal membrane oxygenation.

Thirty two patients (86.5%) were treated with antibiotics, 21 (56.8%) with corticosteroids, 8 (21.6%) with antifungals, 6 (16.2%) with cyclophosphamid, 6 (16.2%) with antivirals, 2 (5.4%) with plasmapheresis and 1 (2.7%) with rituximab.

Fourteen (37.8%) patients were discharged from the ICU: 4 small vessel vasculitis (ANCA+); 3 silicosis; 2 sarcoidosis; 2 COP; 1 fibrotic unclassifiable ILD; 1 RL and 1 scleroderma lung.

Twenty three 23 patients (62.2%) died in ICU: IPF (5); chronic hypersensitivity pneumonitis (4); fibrotic unclassifiable ILD (4); silicosis (2); acute interstitial pneumonitis (2); unclear ILD (2); sarcoidosis (1); small vessel vasculitis (ANCA +) (1); RL (1); dermatomyositis (1). Short term mortality (first month) and overall hospital mortality were 50% (7 patients) and 86.5% (32 patients) respectively.

Four patients did not require IMV: 1 had COP, 1 unclassifiable ILD, 1 small vessel vasculitis (ANCA+) and 1 RL. From this set of patients only the one with small vessel vasculitis (ANCA+) remains alive, the others died in hospital after ICU discharge.

A fibrotic ILD with traction bronchiectasis and honeycombing was associated with a worse outcome ($p = 0.031$ and $p = 0.031$), as well as the need of IMV ($p = 0.007$). A worse outcome was not associated with a higher APACHE II score ($p = 0.198$), a higher SAPS II score ($p = 0.713$), previous oxygen therapy ($p = 0.353$) or previous immunosuppressant therapy (0.982). None of the other parameters analyzed were associated with a worse outcome probably due to the small sample size (Table 1).

We found a high ICU and short term mortality rate, 62.2% and 50% respectively. These findings are consistent with the available literature, which indicates that progression and exacerbation of chronic ILD generally denotes a poor outcome once IMV has been instituted.[4,5]

Fibrotic ILD with CT scan evidence of fibrosis, defined as traction bronchiectasis and honeycombing was associated with a poor outcome. Traction bronchiectasis and honeycombing indicate advanced histopathological alterations, associated with increased lung stiffness, poorer alveolar-capillary gas exchange and greater vulnerability of the lung if IMV is used.[3]

Table 1 ICU and short term outcomes.

	ICU		*P* value	1 month		*P* value
	Nonsurvivors *n* = 23	Survivors *n* = 14		Nonsurvivors *n* = 7	Survivors *n* = 7	
Radiology						
Honeycombing	15/65.2%	4/28.6%	0.032	1/14.3%	6/85.7%	0.237
Traction bronchiectasis	15/65.2%	4/28.6%	0.032	1/14.3%	6/85.7%	0.237
Scores						
APACHE II	19.3 ± 12.3	16.5 ± 5	0.198	14.3 ± 3.2	18.9 ± 7.3	0.352
SAPS	36.1 ± 12.3	38.4 ± 10.2	0.713	39 ± 6.9	36.6 ± 12.1	0.476
NIV	5/21.7%	4/28.6%	0.639	2/28.6%	5/71.4%	1
IMV	23/100%	10/28.6%	0.007	5/71.4%	2/28.6%	1

There was a low rate of lung biopsy, probably reflecting concern about the risk of increased morbidity and mortality associated with this procedure.

The institution of IMV in these patients may raise philosophical questions and would be a procedure to debate, with individualized indications, mostly in cases where the disease is not very advanced or in lung transplant candidates. Physicians should discuss with patients and their relatives the level and degree of life support in case of clinical worsening and ICU admission with IMV should be needed.

Conflicts of interest

The authors have no conflicts of interest to declare.

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A.F. Gonçalves[a,*], S. Campainha[b], C. Nogueira[c], F. Costa[d], P. Castelões[c], S. Neves[b]

[a] *Centro Hospitalar Vila Nova de Gaia/E, EPE, Vila Nova de Gaia, Portugal*
[b] *Centro Hospitalar Vila Nova de Gaia – Pulmonology Department, Portugal*
[c] *Centro Hospitalar Vila Nova de Gaia – Intensive Unit Care, Portugal*
[d] *Centro Hospitalar Vila Nova de Gaia – Radiology Department, Portugal*

* Corresponding author.
E-mail address: ana_f_goncalves@hotmail.com (A.F. Gonçalves).

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Integrated domiciliary ventilation outpatient clinic – Description and experience of an integrated and multidisciplinary model

Long-term domiciliary ventilation has been a growing therapeutic option in children, namely in children with special and complex health care needs. For this reason, multidisciplinar outpatient clinics became necessary. Authors' goal was to describe the organizational model of an integrated domiciliary ventilation paediatric outpatient clinic (IDVC), describing its first 6 years of experience with a retrospective methodology.

Integrated Domiciliary Ventilation Outpatient Clinic was created in 2009 at *Centro Hospitalar do Porto's* Paediatric Department in Portugal. A paediatric pulmonologist (IDVC coordinator), a nurse and a physiotherapist with specialize education in paediatric pulmonology have been the core IDVC working group. Starting in the hospital evaluation, general patient assessment is done (in coordination with various medical specialities, optimizing hospital visits), respiratory monitoring support equipment is adjusted and caregivers are trained in the needed domiciliary care. Since 2012 it is also possible to start long-term ventilation in the outpatient setting, followed by a close monitoring at home. Individual follow-up care plan is performed in coordination with institutions available near home in permanent cooperation with the domiciliary team. This team is formed by health professionals trained in respiratory problems and is coordinated by IDVC physiotherapist. They perform periodic and urgent assessments at home, providing expert technical support and monitoring respiratory therapies, which strengthens therapeutic adhesion and enables a rapid and efficient approach to exacerbations and ventilation problems. The physiotherapist presence at the hospital evaluation allows integration of technical and monitoring data with the clinical assessment leading to a rapid and dynamic flow of information.

Between 2009 and 2014, 93 paediatric patients were followed at IDVC and ventilation initiated in 80 patients (9–15 new patients/year) – Fig. 1. The majority were male (58%) with neuromuscular disease (35.5%), malformative syndrome (24.7%) or cerebral palsy (20.4%). Noninvasive ventilation (NIV) was used in 95% of patients. Ventilation had palliative goal in 90.3%, was initiated at 8.1 ± 6 years

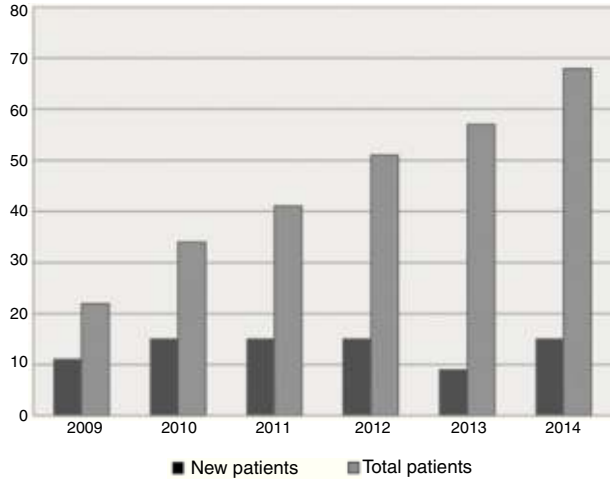

Figure 1 New and total IDVC patients by year, since 2009 to 2014.