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AZTREONAM LYSINE INHALATION SOLUTION (AZLI, CAYSTON®, GILEAD) STABILISES LUNG FUNCTION DECLINE IN PATIENTS WITH CYSTIC FIBROSIS AND MODERATE TO SEVERE LUNG DISEASE

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ABSTRACT BODY:
Introduction: P. aeruginosa (PA) is the most common pathogen and is associated with worse lung function, nutrition and increased mortality. Aztreonam lysine inhalation solution (AZLI, Cayston®, Gilead) is licenced for use in patients with CF aged six years and older, with chronic PA (CPA). The aim of this audit was to assess the usage and clinical outcome of AZLI in a large adult CF unit.
Methods: Data was extracted between 2011 and 2017. A total of 168 patients received at least one dose of AZLI. Indications included CPA, PA eradication and combination therapy in acute exacerbations. Patients who received at least 50% of doses (91/168 patients) were included with 15 subjects being excluded due to pregnancy, lung transplant and recent treatment. Data was extracted for age, weight, BMI, lung function, CRP, plasma viscosity (PV), white cell count (WCC), and days of IVs .
Results: A total of 76 patients met inclusion criteria; PA (n=62) and Burkholderia Cepacia Complex (BCC) +/- PA (n=14). Of these 76 patients, 3 died within 1 year of prescription of AZLI, 4 died within 2 years, and 6 died within 3 years. Prior to the initiation of AZLI, the majority of patients had failed to respond to alternative inhaled antipseudomonal treatment; promixin monotherapy (PA n=17; BCC/PA n=2), tobramycin monotherapy (PA n=28; BCC/PA n=8), or a combination of the 2 treatments (PA n= 14; BCC/PA n=2). AZLI was prescribed on an alternate month basis with another agent in 49 PA (20 promixin, 29 tobramycin) and 8 BCC patients (3 promixin, 5 tobramycin). There were no differences in clinical characteristics at baseline between PA and BCC patients except for weight where BCC patients were heavier. AZLI therapy was initiated in patients with low FEV1. In both groups there was a significant decline in FEV1 prior to initiating therapy ($p<0.001$) which subsequently stabilised (NS). There was a significant increase in CRP and PV in patients with PA over the study period (Table).
Conclusion: AZLI therapy stabilises lung function decline in patients with PA even with moderate and severe disease. Data suggests that AZLI was being prescribed late in the disease course and should be considered sooner. Further work to assess adherence to therapy is needed.
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Table: Within and between group effects (PA v BCC) from 1 year pre-AZLI to 1 year post

Patient characteristic		1 year pre-AZLI	Baseline	1 year post AZLI	Within Group effect	Between Group effect
Weight (Kg)	PA BCC	62.7(±11.9) 68.8(±12.6)	62.9(±12.0) 68.1(±13.1)	62.8(±12.2) 68.5(±13.7)	p = 0.88 p = 0.82	p = 0.62
BMI (Kg/m^2)	PA BCC	22.0(±3.0) 23.6(±3.0)	22.1(±3.1) 23.4(±3.1)	22.1(±3.1) 23.5(±3.1)	p = 0.42 p = 0.83	p = 0.58
FEV_1 (%)	PA BCC	46.2(±14.7) 50.4(±15.1)	42.6(±14.2) 46.8(±15.5)	40.7(±14.8) 43.9(±15.1)	**p < 0.001** **p = 0.008**	p = 0.82
FVC (%)	PA BCC	66.1(±17.4) 72.5(±16.3)	62.8(±16.3) 68.5(±17.8)	60.6(±18.6) 65.4(±18.3)	**p < 0.001** **p = 0.02**	p = 0.62
CRP	PA BCC	16.6(±10.6) 19.8(±17.2)	22.4(±17.9) 15.4(±9.4)	24.5(±20.0) 25.9(±26.4)	**p = 0.005** p = 0.23	p = 0.17
PV	PA BCC	1.8(±0.13) 1.83(±0.11)	1.83(±0.17) 1.84(±0.08)	1.85(±0.19) 1.85(±0.11)	**p = 0.004** p = 0.89	p = 0.37
WCC	PA BCC	6.7(±2.2) 7.7(±2.8)	6.8(±2.1) 7.6(±2.3)	7.1(±2.5) 7.9(±3.0)	p = 0.08 p = 0.79	p = 0.73
Days of IVs	PA BCC	13.9(±7.0) 6.8(±11.5)	15.5(±7.6) 20.3(±21.4)	17.1(±13.2) 14.1(±7.7)	p = 0.09 p = 0.36	p = 0.07