TREATMENT DECISIONS ACCORDING TO 1-YEAR RISK MORTALITY IN PULMONARY ARTERIAL HYPERTENSION PATIENTS: A MULTICENTRE RETROSPECTIVE STUDY

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Background and Importance The 2015 and 2022 ESC/ERS Guidelines for pulmonary hypertension treatment provide algorithms for decision-making based on patients’ 1-year mortality risk, with strong recommendations to intensify treatment in patients with intermediate-high risk.

Aim and Objectives To assess whether treatment decisions in pulmonary arterial hypertension [PAH] patients are currently being made according to the treatment algorithms provided by the ESC/ERS Guidelines.

Material and Methods A retrospective, descriptive, cross-sectional (March 2022) study was carried out in 2 tertiary hospitals, including alive adult PAH patients who initiated a PAH-specific therapy after 2016 and whose medical charts provided enough data to estimate the risk of 1-year mortality with the simplified four-strata risk-assessment tool.

Medical charts were consulted in order to collect several variables: demographic data, PAH subclassification according to aetiology, PH-specific drug initiated, World Health Organization functional class [WHO-FC], 6-minute walking distance [6MWD], and N-terminal pro-brain natriuretic peptide [NT-proBNP].

1-year mortality risk and the appropriateness of PH-specific therapies prescribed were assessed according to PAH treatment algorithms provided by the 2015 and 2022 ESC/ERS Guidelines.

Results 37 patients complied with inclusion criteria, 54.1% women aged 50 (28–84).

Patients’ HAP subsets: 14, 6, 2, 2, and 1 were associated with adult congenital heart disease, portal hypertension, connective tissue disease, drugs and toxins, and human immunodeficiency virus infection, respectively. 6 patients were classified as idiopathic HAP.

52 changes in pulmonary-specific therapy were carried out in the studied period. At treatment initiation patients:

- WHO-FC: I, II, III, and IV in 2, 21, 26, and 3 cases, respectively.
- 6MWD: 425 (146–760) metres
- NT-proBNP: 369 (12–7200) ng/L
- Risk: 17 low; 20 intermediate-low; 14 intermediate-high, and 1 high.

36/52 treatment initiations were adequate according to clinical guideline algorithms; most discrepancies were due to:
- Initiation of selexipag (n=9) or riociguat (n=3) in patients with risk other than intermediate-low.

Conclusion and Relevance In this cohort of PAH patients whose 1-year mortality risk could be estimated, treatment decisions were generally made according to treatment guidelines.

Patients’ preferences could explain most discrepancies, as they may prioritise avoiding treatments that require parenteral administration, such as epoprostenol and treprostinil and rather try oral alternatives.

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the mortality risk is being assessed but not registered in clinical charts.

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6ER-014 EFFECTIVENESS AND SAFETY OF THE ADMINISTRATION OF MURPHY’S ENEMA FOR THE TREATMENT OF REFRACTORY CONSTIPATION IN A TERTIARY HOSPITAL
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Background and Importance Constipation is a common complication during hospitalisation due to the presence of risk factors such as bed rest, diseases causing reduced bowel motility or administration of medications (opioids, anticholinergic drugs...). The standard therapy is laxative drugs. Murphy’s enema (ME) is used for the treatment of constipation and faecal impaction when patients do not respond to laxatives. It consists of administering an evacuating solution (milk 300 mL, liquid vaseline 100 mL, oxygenated water 200 mL and saline solution 500 mL) through a rectal probe during 6 hours (53 drops/min), leading to a softening of the stool and osmotic evacuation. Although this is a common clinical practice in our hospital, we have not found any published study evaluating its effectiveness and safety.

Aim and Objectives To assess the effectiveness and safety of ME for the treatment of constipation and faecal impaction.

Material and Methods We performed a descriptive, retrospective study of effectiveness and safety of the administration of ME in patients with admitted in a tertiary hospital. We included patients who received ME from June-2020 to August-2022. We registered data of comorbidities, defecation achievement and adverse events related to the administration. Data were obtained from the electronic prescription program and the electronic health records.

Results We included 33 patients, 18 women and 15 men, with a mean age of 76 years. Two patients were readmitted, therefore a total of 37 ME were administrated. The most frequent comorbidities were hypertension (40,5%), chronic constipation (33,3%), diabetes mellitus II (21,2%), heart failure (18,1%), atrial fibrillation (18,1%), dyslipidemia (12,1%), cognitive impairment (12,1%) and kidney failure (12,1%). The indication of ME was constipation (67,5%), faecal impaction (27,2%) and paralytic ileum (5,4%). ME was effective in the 64,8% of cases, with defecation achievement after administration. ME was well tolerated in all patients; one case of hypotension, one of nausea and one of abdominal pain were registered.

Conclusion and Relevance ME constitutes a safe and effective alternative for patients with constipation and faecal impaction not responding to the usual therapies. Furthermore, there is no published evidence regarding this practice, so this study may constitute a starting point for the development of further studies with larger sample sizes.

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6ER-015 AZACITIDINE IN THE TREATMENT OF JUVENILE MYELOMONOCYTIC LEUKAEMIA: AN UP-TO-DATE PHARMACY PROTOCOL
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Background and Importance Juvenile Myelomonocytic Leukaemia (JMML) is a paediatric haematological malignancy with a poor prognosis. In August 2019 in our paediatric hospital, we have a case of JMML. A protocol pharmacy related to medicinal product indication, preparation, and flow chart instructions was made based on the information given by the patient’s doctor, marketing authorisation, and internet research. In Europe, azacitidine is indicated for the treatment of adult patients who are not eligible for haematopoietic stem cell transplantation (HSCT) with specific diagnostic criteria. The reconstituted solution should be injected subcutaneously. For this reason, diagnosis and route of administration, this request was an off-label use. Azacitidine is cytotoxic and is prepared in a centralised production unit, under the pharmacist’s responsibility. In 2022 we intended to research the current state of the art.

Aim and Objectives A systematic review of azacitidine in the treatment of JMML, in a recent period of time and, based on the results, update the pharmacy protocol of our hospital.

Material and Methods To perform this work, we used the following databases: PubMed and Embase, limited to publication years from 01 January 2020 to 09 September 2022. Key words included: azacitidine AND Juvenile Myelomonocytic Leukemia. An Excel table was made with the results.

Results Data synthesis: We found 57 articles. Among them, 27 were excluded by the title and 10 by the summary. Among the 20 analysed manuscripts, 6 were repeated and 8 were excluded after reading the full text. Thus, 6 articles were selected for this review.

Conclusion and Relevance A significant change occurred in May 2022. Food and Drug Administration (FDA) approved azacitidine monotherapy as a suitable option for children with newly diagnosed JMML based on the results of the AZA-JMML-001 trial. Although long-term safety and efficacy remain to be fully elucidated in this population, the data demonstrate that azacitidine provides valuable clinical benefit to JMML patients prior to HSCT. In Europe, it has not yet been approved for this clinical situation.

It is important to share treatments for rare diseases. Pharmacists are medication experts and play a critical role in this. Accordingly, we review a pharmacy protocol and update azacitidine new findings.

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