Residência RP Pediátrica
Publicação Oficial da Sociedade Brasileira de Pediatria

Submitted on: 06/07/2019
Approved on: 06/11/2019

POINT OF VIEW

Pediatric rheumatology and palliative care for children: a most relevant match

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Keywords:
Palliative Care,
Child,
Adolescent,
Rheumatology,
Pain,
Quality of life.

Abstract

The overall profile of pediatric patients has changed in recent years, as the proportion of children with chronic or life-threatening conditions in need of care has increased. In this setting, pediatric palliative care has emerged as a means to provide integral care to patients and their families. Pediatric rheumatology is closely tied to palliative care for children, since patients seen under this medical specialty are often diagnosed with severe, life-threatening diseases. This study aimed to describe the intersections between the two medical specialties in current practice. At first we thought of writing a literature review, but to our surprise we were unable to find papers specifically discussing the connections between pediatric rheumatology and palliative care for children. Despite the lack of literature on the subject, there is growing concern among pediatric rheumatologists over the wellbeing of pediatric patients and their families. The ideas of palliative care must be systematically extended to pediatric rheumatology, since lack of knowledge on the matter impedes the adoption of broader and more adequate approaches. The severity and morbidity of pediatric rheumatic diseases call for the early introduction of palliative care through the hands of specifically trained teams. This study sheds light on the urgent need to introduce notions of palliative care to health care providers involved in pediatric rheumatology.

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Residência Pediátrica 2019;9(2):189-192. DOI: 10.25060/residpediatr-2019.v9n2-21

INTRODUCTION

The overall profile of pediatric patients has changed in recent years, as the proportion of children with chronic or life-threatening conditions in need of care has increased. In this setting, pediatric palliative care (PPC) has emerged as a means to provide integral care to patients and their families.

PPC plays a key role in providing relief and treating the signs and symptoms of disease via interventions in a number of areas, including psychological and spiritual support. In order to help patients cope with their fate and families grieve, death is seen as part of a natural process, as something that should not be rushed or prolonged, while life is celebrated. Support networks are formed so that patients can live as actively as possible and families may learn to adjust to the situations imposed on them by the disease, having teamwork as a guiding principle[1].

Different pediatric conditions may benefit from palliative care. In order to clarify indications, meet patient needs, and leave no child without access to integral care, patients are divided into four groups, as follows[2]:

Group 1 - Potentially life-threatening conditions in which cure is a possibility, including diseases such as cancer and sepsis.

Group 2 - Conditions requiring long-term intensive care to prolong life with an ever-present chance of premature death. Diseases such as cystic fibrosis, epidermolysis bullosa, muscular dystrophy, sickle-cell anemia, and severe cardiovascular anomalies belong in this group.

Group 3 - Progressive diseases without a cure, in which palliative care is instituted from the time of diagnosis. Examples include metabolic disorders and chromosomal abnormalities.

Group 4 - This group includes irreversible non-progressive diseases, in which patients face severe disabilities that increase vulnerability to complications. Examples include genetic diseases, malformations of the central nervous system, and cerebral palsy.

A large number of diseases may benefit from PPC, most with variable and unpredictable treatment duration[2]. This is the reality of practice in pediatric rheumatology (PR), in which physicians often face the daunting task of predicting the degree of disability and the risk of death of patients with rheumatic conditions despite the availability of advanced therapies[3].

A significant portion of rheumatic diseases affects pediatric patients[3]. They are difficult to manage, and severe forms may be life threatening. Significant progress has been made in PR, particularly after immunobiologicals were discovered. Although cure might not be a possibility for some patients, many children diagnosed with untreatable conditions in the past may be partially treated today. However, they are often forced to live with marked - and often incapacitating - sequelae derived from the disease or prescribed medication.

Assuming that palliative care does not exclude curative therapy and that focus resides on improving quality of life[1], it is our belief that PPC and PR should walk hand in hand.

OBJECTIVE

As we thought about PR and PPC, it only seemed natural that the two were intertwined, since the first revolves around providing care to patients with severe, life-threatening conditions and the second brings in a more holistic approach to care. Therefore, this study aimed to show the intersection between the two medical specialties in current practice.

METHOD

At first we thought of writing a literature review, which prompted us to search for papers in Brazilian and international databases, namely the Biblioteca Virtual em Saúde (which includes Lilacs e Scielo) and PubMed.

The following keywords were used: (Reumatologia OR Rheumatology OR Rheumatic Diseases OR Rheumatism) AND (Cuidado Paliativo OR Palliative Care OR Palliative Medicine OR Palliative Care Medicine).

The search included papers published within an undetermined period of time, which would later be processed and analyzed to assess for possible matches with the subject of pediatrics.

RESULTS

To our surprise, we were unable to find papers specifically discussing the connections between pediatric rheumatology and palliative care for children in the searched databases.

The search on Biblioteca Virtual em Saúde yielded one paper, but it had nothing in connection with palliative care and rheumatology.

The 120 papers found on PubMed were individually analyzed. Only one versed specifically on the combination of rheumatology and palliative care, but it made no reference to pediatric populations.

DISCUSSION

The tenets of palliative care have somehow been partially incorporated in PR. Despite the lack of literature on the subject, many health care providers strive to improve the wellbeing of pediatric patients and their families. However, broader and more adequate approaches can only be implemented after PPC has been

systematically introduced in the practice of pediatric rheumatology.

Many children with rheumatic diseases are eligible for palliative care based on the criteria presented in the Introduction section. Patients with certain subtypes of juvenile idiopathic arthritis (JIA) fit into Group 1, since this is a life-threatening, possibly curable condition depending on the type of involvement and pattern of disease progression. Group 2 may very well include patients with juvenile-onset systemic lupus erythematosus (SLE) and Takayasu arteritis, which require long-term treatment and pose a risk of premature death. Group 3 may include children with advanced dermatomyositis, since it is a progressive disease for which there is no cure in some cases. Patients with stroke secondary to antiphospholipid antibody syndrome (APS) have possibly irreversible sequelae and may be included in Group 4 on account of severe disability.

PPC and PR teams can work together, since the treatments provided by both are mutually non-exclusive. Interdisciplinary care is common in PR, as cardiologists and physicians from other specialties are often involved in patient care. Multidisciplinary care is also provided routinely, as physiotherapists, nurses, psychologists, and social workers often engage with patients. The point is that most of the time patients are seen by individual health care workers and not by an integrated team. In PPC, patients are supposed to be engaged by a team of health care workers from various disciplines who meet periodically to discuss care measures and seek to continuously interact with and provide tailored care to patients and their families.

Diagnosing and managing pain is one of the pillars of symptom management in PPC[1,4], given that pain is the most common symptom in patients with rheumatic disease.

Although patients with JIA are often on anti-inflammatory medication, disease-modifying antirheumatic drugs (DMARDs), and immunobiologicals, pain frequently persists. Given its physical and emotional components[1], pain is more satisfactorily treated with multidisciplinary care.

Pediatric rheumatologists must extend their knowledge of how to diagnose pain, learn additional pain categorization schemes and scales, find out more about potential therapies, and master the principles of the WHO analgesic ladder. The expertise held by PPC teams is greatly needed in the management of more specific cases requiring comprehensive care strategies. Integrative medicine may have a role to play here, since many of these patients are on multiple drugs. Complementary non-drug therapies such as acupuncture and Reiki[5] may be added. Physical practices and physical activity are a part of treatment to be considered by health care teams[5].

Patients must be periodically monitored by a pediatric rheumatologist and the PPC team depending on the severity of the disease and the particularities of each patient. Having a second pediatrician to assess patient growth and development, help to manage symptoms, provide encouragement, and bond with the patient is of paramount importance. A physiotherapist can monitor patient motor development and deal with possible physical and respiratory impairment. A physical educator can encourage patients to engage in sports activities and adjust them when needed. A nutritionist can expertly follow children with chronic diseases. An occupational therapist can help patients develop skills and integrate. A nurse can provide specific care to patients with acute or chronic conditions. A dentist can assist in oral care, given that many diseases present with recurrent oral involvement. A pharmacist can provide advice on which medication is suitable to each individual patient. Spiritual and psychological counseling gain additional relevance for patients and their families, as current therapies may be insufficient and produce complex physical and emotional sequelae. Social workers play a pivotal role in PPC teams, as they help families cope with the challenges imposed by multiple medical appointments and hospitalizations, medication purchases, and other issues affecting the ability of family members staying at their jobs.

Difficult issues must be gradually addressed as part of a care plan developed jointly with patients, their families, and the PPC and PR teams[2].

CONCLUSION

The severity and significant morbidity characteristically seen in pediatric rheumatic diseases call for the early introduction of palliative care to preserve the quality of life of patients and their families.

Multi- and interdisciplinary teams with specific training in PPC must be assembled to address the needs of pediatric patients and children with rheumatic diseases in particular. While a few centers have adopted practices from palliative care to treat children with rheumatic diseases, in most healthcare institutions the issue has been given insufficient attention.

This study sheds light on the urgent need to introduce notions of palliative care to health care providers involved in pediatric rheumatology and in managing patients with chronic diseases.

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