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ARTICLE REFLEXIVE ANALYSIS

SPOAN SYNDROME: A MATTER OF CONSANGUINITY AND OF CONQUEST OF RIGHTS

SÍNDROME SPOAN: UMA QUESTÃO DE CONSANGUINIDADE E DE CONQUISTA DE DIREITOS SÍNDROME SPOAN: UNA CUESTIÓN DE CONSANGUINIDAD Y DE CONQUISTA DE DERECHOS

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ABSTRACT

Objectives: to reflect on the influence of consanguineous marriages in determining SPOAN syndrome and health care offered to patients with the syndrome in view of the National Health Policy of Persons with Disabilities. **Method:** reflective study based on literature review. **Results:** SPOAN syndrome, affects the central nervous system. Features progressive evolution and why carriers need assistance that prioritizes quality of life, for that the National Health Policy of Persons with Disabilities (PNSPPD) must be present in the construction of citizenship of these subjects. **Conclusion:** The National Health Policy of Persons with Disabilities should underpin health actions undertaken by local authorities, to be effective in improving the quality of life of patients. **Descriptors:** SPOAN; Optic atrophy; Hereditary Spastic Paraplegia.

RESUMO

Objetivos: refletir sobre a influência dos casamentos consanguíneos na determinação da Síndrome SPOAN e a assistência à saúde oferecida aos portadores da síndrome tendo em vista a Política Nacional de Saúde da Pessoa Portadora de Deficiência. **Método:** estudo reflexivo baseado em revisão de literatura. **Resultados:** a Síndrome SPOAN, acomete o sistema Nervoso Central. Apresenta evolução progressiva e por isso os portadores necessitam de uma assistência que priorize a qualidade de vida, para isso a Política Nacional de Saúde da Pessoa Portadora de Deficiência (PNSPPD) deve fazer-se presente na construção da cidadania desses sujeitos. **Conclusão:** a Política Nacional de Saúde da Pessoa Portadora de Deficiência deve nortear as ações em saúde desenvolvidas pelo poder local, para que sejam efetivas na promoção da qualidade de vida dos pacientes. **Descritores:** SPOAN; Atrofia óptica; Paraplegia Espástica Hereditária.

RESUMEN

Objetivos: reflexionar sobre la influencia de los matrimonios consanguíneos en la determinación de síndrome Spoan y la atención sanitaria ofrecida a los pacientes con el síndrome de visión de la Política Nacional de Salud de las Personas con Discapacidad. **Método:** estudio reflexivo sobre la base de revisión de la literatura. **Resultados:** Síndrome Spoan, afecta al sistema nervioso central. Características evolución progresiva y por qué las compañías necesitan asistencia que da prioridad a la calidad de vida, para que la Política Nacional de Salud de las Personas con Discapacidad (PNSPPD) debe estar presente en la construcción de la ciudadanía de estos temas. **Conclusión:** la Política Nacional de Salud de las Personas con Discapacidad debe sustentarse en acciones de salud llevadas a cabo por las autoridades locales, para ser eficaz en la mejora de la calidad de vida de los pacientes. **Descriptores:** Spoan; Atrofia óptica; Paraplejia Espástica Hereditaria.

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INTRODUCTION

In 2005, researchers at the Center for Human Genome Studies and Hospital das Clínicas, University of São Paulo (USP) have discovered a new neurodegenerative disease and described in an article published in the U.S. journal *Annals of Neurology*. The disease was called autosomal recessive syndrome SPOAN (Spastic Paraplegia acronym, Optic atrophy, and Neuropathy) whose initial characterization was performed in individuals originating from Serrinha Pintos located on top RN west.

It is estimated that one in every seven neighbors of the municipality is a carrier of the disease associated mutation in heterozygous state. Serrinha is among the 50 municipalities of Brazil with the highest rate of individuals with disabilities, and certainly the high prevalence of the syndrome SPOAN contributes to that.¹

Recognizing the high prevalence of individuals with disabilities in high-west, with particular attention to the recent discovery of the syndrome in SPOAN macroregion where we live, and that the law guarantees access to health promotion, medical differentiated, home visits and specific health programs so that people with disabilities have access to the network so integral, associated with the fact that during the curricular supervised held in the upper west RN, the same region of the city of Serrinha Pintos, we encounter a patient with SPOAN syndrome, we observed the paucity of sources about the topic and little knowledge on the part of academics and health professionals despite the high prevalence of cases of the syndrome in the region.

On visits to the municipality noticed how poor the assistance is targeted to those with SPOAN, motivating us to reflect on the influence of consanguineous marriages in determining SPOAN syndrome and health care offered to patients with the syndrome in view of the National Health Policy of Persons with Disabilities / PNSPPD.

• SPOAN: a familiar case

It is said in Serrinha Pintos in Rio Grande do Norte, Northeastern Brazil, it all started with Maximiniano that came to town to marry Antonia, daughter of Pedro Queiroz. The old womanizer, who contracted syphilis, once he found himself a widower, married a niece of Antonia. From there, making a period of about 200 years, it had settled syphilis blood family, which grew into a town where all are, to a greater or lesser degree relatives.²

This syndrome was Recognized Already in this region by the end of 19th century and at least six more Individuals were reportedly affected. Interestingly, the concept that this condition is genetically determined was not present in the community, and several other Reasons were evoked to explain this phenomenon, most of them related to syphilis.^{3:731}

When making an argument about staying still, syncretic explanations for the natural and social phenomena even in a period when there is spread of education, mentions the tradition of consanguineous marriage of Serrinha Pintos and says that, like any other disease who is transmitting a recessive manner, as is the case of the new entity, the frequency of individuals affected with such practice increases.⁴

By analyzing what they say about the city on the "Old Maximiniano" noted that "the narrative not only explains the origin and inheritance of the disease but also to deny their relationship with inbreeding, which enables people to maintain and enhance these practical wedding consanguineous".^{4:8}

Looking at the family tree, biologist researcher Silvana Cristina dos Santos said that the disease began in the marriage of Peter and Mary Alexandrina Queiroz, parents of Antonia, wife of Maximian. The couple had three children, who had the defective gene. When her grandchildren and great-grandchildren started marrying among themselves, the disease appeared. Syphilis old Maximiniano therefore not associated with their emergence. What happens is that between the sixteenth and nineteenth centuries it was believed that syphilis was linked to hereditary evils and even today this belief is present between the families of Serrinha Pintos.²

Consanguineous marriages have always been a local habit and such marriages represent a higher risk to generate individuals with disease. Beiguelman mentions that, among individuals affected by autosomal recessive diseases, the percentage of those who are children of consanguineous couples can reach very high figures, since such couples are more likely to produce homozygous children than non-kin.⁵

In 2005, Macedo-Souza and colleagues at the Center for Human Genome Studies and the Hospital das Clínicas, University of São Paulo (USP) described in an article published in the U.S. journal *Annals of Neurology* syndrome Serrinha and characterized as a complicated Hereditary Spastic Paraplegia form (PEH) previously unknown.³ A new clinical condition was described in a large family with a huge

Oliveira LC, Vasconcelos RB de, Fixina EB.

Spoan Syndrome: a matter of consanguinity...

number of consanguineous marriages, with 25 affected individuals, 22 of whom were evaluated.

The term "hereditary spastic paraplegia" or "Hereditary Spastic Paraparesis" was described in 1880 used to describe a group of inherited neurodegenerative diseases.

Hereditary Spastic Paraplegia (PEH) is not a single disease but a group of clinically and genetically different disorders characterized by muscle weakness and spasticity bilateral symmetric insidious progression starting with the lower limbs.⁶

PEH is classified as "simple" (weakness of the lower limbs, urinary urgency and decreased vibration sensation in the toes) or "complicated" when other neurologic deficits are present.⁶

The SPOAN syndrome, a type of PEH complex affects their carriers in more than one route or region in the central nervous system. In general, the signs that accompany the PEH are complex: spasticity, weakness, hyperreflexia, and Babinski sign, featuring the pure PEH plus neurologic or non-neurologic, including motor neuropathy and peripheral, optic atrophy, mental retardation, dementia, among others.

About PEHs was observed:⁷

The age of onset of HSP varies from infancy to the eighth decade. The prevalence of HSP was found to be between 2.0 to 9.6/100000 [...]. Autosomal dominant inheritance of HSP is the most common mode of HSP and responsible for about 80% of HSP cases.^{8:571}

Several molecular studies were performed in order to pinpoint the mechanisms involved in degeneration that characterizes the PEH, without, however, succeed. Some hypotheses have been identified. The explanation could come from the lack of production of adenosine triphosphate (ATP) these fibers by a deficiency of mitochondria and other organelles. The genes responsible for this process block the transport of these substances along the axon and the longer they are more susceptible to this interruption, explaining why the degeneration occurs in specific cell types so. A second hypothesis is abnormal myelination of axons induced by an abnormal intracellular trafficking.⁷

The new discovery in PEH Serrinha Pintos was named SPOAN syndrome. This new entity refers to a syndrome progressive, neurodegenerative autosomal recessive. It was proposed the acronym SPOAN (spastic paraplegia, optic atrophy, and neuropathy) for this complex syndrome. "It is Estimated That 1 in every 170 inhabitants of Serrinha Pintos

(population in 2000: 4,250) is affected by SPOAN."^{9:4}

As the vast majority of other PEHs recessive, not yet know which gene is the mutation that causes the disease. An analysis of samples of Deoxyribonucleic Acid (DNA) of the city residents points to the 11q13 region. Until today, none were found associated with neurological disease genes in this region of chromosome 11.⁹ However, a PEH also complex, but autosomal dominant syndrome called Silver, was identified in the region 11q12-14, next to that candidate to determine SPOAN syndrome.

Despite having located the region where the gene responsible for the syndrome, it contains at least 143 genes, of which 96 are activated in nervous tissue. Stated differently it is necessary to find a specific gene within almost one hundred.¹⁰

Clinically, SPOAN syndrome is characterized by:

1. Non-progressive congenital optic atrophy; 2. spastic paraplegia with onset in infancy; and 3. juvenile progressive motor and sensory axonal neuropathy. Additional common findings were dysarthria, spine deformity, and joint retractions, as well as startle response following unexpected noises.^{11:382}

The most frequent clinical feature in individuals affected by congenital SPOAN and atrophy of the optic nerve, already present in the first months of life, and apparently does not progress. The fixed nystagmus, abnormal involuntary movements of the eyeball, present at birth, reduces the field of vision of patients.³

Symptoms related to optic atrophy were recognized early in life and apparently were not progressive. Fixation nystagmus was observed in 18 patients (82%) and was caused by subnormal vision, which was seen in 21 of 22 patients (95.5%) who had pale optic disks.^{3:731}

Signs of onset of motor deficits usually appear before a year. Abnormalities of motor development and progressive spastic paraplegia of the lower limbs take the patient to develop a march held on tiptoe. In most cases, these abnormalities do not impede the patients, even early frail, unable to get around until around age 10, when this march is then lost. The use of aid as walkers and canes are usually used until 20 years old, then become insufficient when making use of the wheelchair is essential. The involvement of the lower limbs always precedes the above, and usually the deficiency of the former is more intense.¹⁰

Some townspeople say that some affected children are born through "molinhas" apparently unsteadily to sit or even crawl. But even these fragile patients early can usually get around on their own or supported with frequent falls and tropicans until a certain age, usually around 10 years...^{10:2}

Regarding the installation of motor and sensory neuropathy usually perceives the occurrence of distal muscular atrophy, occurring most often in patients older than 20 years and not notice any sign of fasciculation. There is an overlap of signals and peripheral pyramidal and thus reflections are obtained more easily proximal to the distal. Hyperhidrosis, changes in tactile sensitivity, and lack of sensitivity deep distal signs are common among people affected by the syndrome.³ Complements to the concept that even in later stages there is no change in the perception of pain and temperature.

Another feature very common among patients with dysarthria SPOAN is associated with low tone also affects individuals older than 20 years. Such disorders are sometimes so intense to the point of compromising speech understanding for some patients.⁷

An abrupt motor response when they are taken by surprise by an unexpected sound is a peculiarity seen in these patients. Interesting that even those who did not outline any voluntary movements in the lower limbs, exhibit this type of involuntary muscle contractions in these conditions. This event is present in all patients with SPOAN observed even after adulthood, being easily provoked, including severe cases and advanced disease.³

All examined patients presenting varying degrees of limitation have deformities in the spine and joints, associated with limited mobility especially of the ankle, knee, wrist and elbow. Varying degrees of cervicothoracic kyphosis and scoliosis are very common features among these patients may reach degrees so severe that even disabling individual to sit independently.³

The intensity of the clinical picture may vary among patients, however the disease is quite insightful, and its effects on the quality of life of those affected by the syndrome even be devastating. Nevertheless, patients do not exhibit cognitive impairment, or mental retardation, ataxia or deafness not part therefore of clinical symptoms of the syndrome SPOAN.¹⁰

It is important to mention that the combination of PEH with axonal neuropathy, dysarthria and congenital optic nerve atrophy as observed in patients with the syndrome in

Serrinha Pintos, has never been described previously.³

The PEHs with autosomal recessive inheritance is present in the vast majority as a complex form. In Dillmann et al describes a study with two affected in an inbred family that housed the association since childhood progressive spasticity, peripheral neuropathy and progressive optic atrophy started in adolescence. The disease reported by Dillmann et al also showed an autosomal recessive pattern as in SPOAN syndrome.¹²

The difference between the two bodies is given by the later onset of optic atrophy and the insidious onset of spasticity of the PEH.

A recent study sought to evaluate motor performance and functional SPOAN included 61 individuals with an age range from 5 to 72 years old. The same revealed that spasticity shows a bimodal distribution, with Grade 1 (minimal) to 4 (maximum). The grip strength of the hand showed an inverse correlation with age. Thus, it was concluded that the combination of spastic paraplegia with early-onset progressive polyneuropathy syndrome makes SPOAN a disabling condition.⁹

In summary, this investigation allows quantify the motor and functional performance of 61 individuals diagnosed with SPOAN and suggests that this complicated form of HSP has a lifelong progression. Specific scales for HSP did not appreciate disease progression, as functional deterioration of lower limbs is an early event and achieves a maximum before 20 years of age.^{9:6}

In PEHs, imaging studies of the brain and spinal cord magnetic resonance imaging (MRI) are generally normal. Only in complicated shapes as is the case SPOAN, the nerve conduction studies on electromyography may show sensory and motor impairment with axonal characteristics of peripheral nerves.

Researchers at the Center for Human Genome Studies and the Hospital of the USP were used MRI, which showed no change, and electromyography revealed that the typical characteristics of the aforementioned PEHs complicated. It was also used in an electroretinogram a study of cerebrospinal fluid that did not show any changes. Some patients had spinal deformity in scoliosis radiography characteristics.

Based on the clinical mentioned herein Pivetta emphasizes that the effects are devastating disease with a major impact on the quality of life of those affected, particularly by affecting a population rural poor and Serrinha such as "believes, so folklore, the origin of a new disease syphilis

Oliveira LC, Vasconcelos RB de, Fixina EB.

Spoan Syndrome: a matter of consanguinity...

inherited from an ancient ancestor and womanizing that would spread through the blood of the family.”^{10:1}

Understanding the relationship between inbreeding and genetic manifestation of the disease is essential for the inhabitants of Serrinha Pintos can make reproductive decisions aimed at preventing the birth of more people affected.¹³

In SPOAN syndrome, symptoms progress over the years and the affected individuals close to their homes living apart from day-to-day, collected, and totally dependent on the care of relatives. Deprived of adequate medical care, no one receives any form of treatment in order to reduce stiffness and weakness of the limbs.¹⁰

According to what was said above people with this syndrome are the margin of dignified care that is already guaranteed by law in Decree 914, and the National Health Policy of Persons with Disabilities, for example.

Provisions of Law No. 7853, Decree 914 of September 6, 1993 establishing the National Policy for the Integration of Persons with Disabilities later regulated by Decree No. 3.298/1999.¹⁴ In summary this policy were established standards regarding health, education, habilitation and rehabilitation, work, culture, sports, tourism, leisure and accessibility.¹⁴ She embodies an instrument which comprises the set of normative guidance that aims to ensure the full exercise of individual and social rights of people with disabilities.¹⁵

The GM / MS 1060, to June 5, 2002, which established the National Health Policy of Persons with Disabilities (PNSPPD) that is directed to include people with disabilities throughout the service network of SUS, press the rehabilitation, protect their health and the prevention of injuries to determine the onset of disability, through the articulation of sectors of society and society itself.¹⁶

Increased already referred this reality when he mentions that despite this progress, the demand is still far from being met. This occurs especially in rural areas and in the poorest regions of Brazil.¹⁷

The researcher Silvana Santos points to the need to concentrate on studies in Serrinha SPOAN disease in order to improve the quality of life of those affected, especially the younger ones who can benefit from physical therapy assistance and maintain good posture.

Studies that account for this necessity must address six main areas: physical health, psychological state, level of independence, social relationships, environmental

characteristics and spiritual standard. To achieve such abstract domains of reality of the disabled are needed instruments or questionnaires that are able to subjectively measure the conditions of well-being of individuals achieving this with a more complete assessment of the impact of the disease and its treatment on patients' lives.¹⁸

CONCLUSION

The National Policy for the Integration of Persons with Disabilities caveat the relevance of some factors of fundamental importance for the promotion of health and quality of life: education, habilitation and rehabilitation, work, culture, sports, tourism, leisure and accessibility and is recognized before the 1988 Federal Constitution how important are these factors to have a quality life. The denial of the standards set by the National Policy for Integration of the Disabled, either relative or absolute, reflected in the quality of life of patients for the services offered to them, although guaranteed by law.

Regarding the health sector, one must have a continuous planning with periodic assessment of the impact of assistance and health care in their lives.

The National Health Policy of Persons with Disabilities should serve as a guide in the planning of activities to be undertaken with the public and patients in the city of SPOAN Serrinha Pintos / RN, as well as the assessments that must be carried out before the tour that is offered to those users.

For the guidelines of this policy are realized, it is necessary the joint action of local government committed and professional team of Doctors and Nursing. Otherwise, practitioners are forced to spend time staring and patients go consciously surrendering to the impositions of the disease and the local socio-economic constraints.

Finally, health professionals who assist users SPOAN carriers must provide him with sufficient information about the syndrome and recognize that people with SPOAN have particularities that put them at odd situation compared to other citizens of Serrinha Pintos.

It should be recognized that the health and other social sectors if they needed to ensure a proper quality of life to these people and that, therefore, it is necessary to develop and implement public policies at the local level that may favor the SPOAN syndrome patients.

To this end, we recognize the need for diagnostic studies in order to visualize the actual needs and thereby promote further discussion, pointing out new horizons for

health professionals responsible for the care of these patients, especially doctors and nurses.

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Oliveira LC, Vasconcelos RB de, Fixina EB.

Spoan Syndrome: a matter of consanguinity...

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