

ORIGINAL ARTICLE

# Emotions at diagnosis in patients with bullous diseases

Francesca SAMPOGNA<sup>1</sup>\*, Silvia BATTISTI<sup>2</sup>, Chiara SCARPULLA<sup>2</sup>, Valentina BATTISTI<sup>2</sup>,  
Federica COSENZA<sup>2</sup>, Carola PULVIRENTI<sup>3</sup>, Giuseppe FORMATO<sup>3</sup>, Damiano ABENI<sup>1</sup>

<sup>1</sup>Clinical Epidemiology Unit, Istituto Dermopatico dell'Immacolata IDI-IRCCS, Rome, Italy; <sup>2</sup>Integrated Psychodynamic Institute (IRPPI), Rome, Italy; <sup>3</sup>Italian National Pemphigoid Patient Organization (ANPPI), Rome, Italy

\*Corresponding author: Francesca Sampogna, Clinical Epidemiology Unit, Istituto Dermopatico dell'Immacolata IDI-IRCCS, Via dei Monti di Creta 104, 00167 Rome, Italy. E-mail: [fg.sampogna@gmail.com](mailto:fg.sampogna@gmail.com)

*This is an open access article distributed under the terms of the Creative Commons CC BY-NC license which allows users to distribute, remix, adapt and build upon the manuscript, as long as this is not done for commercial purposes, the user gives appropriate credits to the original author(s) and the source (with a link to the formal publication through the relevant DOI), provides a link to the license and indicates if changes were made. Full details on the CC BY-NC 4.0 are available at <https://creativecommons.org/licenses/by-nc/4.0/>.*

## ABSTRACT

**BACKGROUND:** Diagnosis of a severe condition may have a strong emotional impact on patients. Specific emotions experienced when receiving the diagnosis of a bullous disease have not been investigated.

**METHODS:** Adult patients diagnosed with a bullous condition were recruited through the Italian Association of patients with pemphigus and pemphigoid (ANPPI). Information was collected online on sociodemographic and clinical data. We asked which emotions the patient experienced at the time of the diagnosis, *i.e.*, isolation, anger, confusion, sadness, despair, disregard, fear, avoidance, and challenge. Also, the patients reported to whom they talked as soon as they had the diagnosis.

**RESULTS:** Data were collected on 105 patients, most of whom were affected by pemphigus vulgaris. The emotion most frequently experienced at diagnosis was confusion (47.6% of patients). More than 30% of patients reported sadness and fear, 12.5% anger, and 10.5% despair. A significantly higher percentage of women than men experienced isolation and despair. Despair was more frequent in patients who were older at diagnosis. Patients with children experienced more sadness and despair, and less avoidance and challenge, while those who had a relative with a bullous disease reported less fear, and more challenge.

**CONCLUSIONS:** Clinicians should be aware of the emotions of the patient when communicating the diagnosis of severe conditions, such as bullous diseases. Active listening and empathy are necessary to provide patients with correct information on the disease, so that they are not overwhelmed with negative emotions.

*(Cite this article as: Sampogna F, Battisti S, Scarpulla C, Battisti V, Cosenza F, Pulvirenti C, et al. Emotions at diagnosis in patients with bullous diseases. Ital J Dermatol Venereol 2023;158:328-33. DOI: 10.23736/S2784-8671.23.07535-7)*

**KEY WORDS:** Vesicobullous skin diseases; Diagnosis; Emotions; Bullous pemphigoid.

Autoimmune bullous diseases are a group of rare, blistering disorders involving the skin and/or mucous membranes.<sup>1</sup> In the pemphigus group (*i.e.*, pemphigus vulgaris, pemphigus foliaceus, paraneoplastic pemphigus, IgA pemphigus, and paraneoplastic pemphigus) autoantibodies target desmosomal proteins that results in loss of cell adherence between keratinocytes. In pemphigoid diseases (*i.e.*, bullous pemphigoid, mucous membrane or cicatricial pemphigoid, linear IgA bullous dermatosis, and epidermolysis bullosa acquisita) hemidesmosomal proteins of the dermo-epidermal junction are targeted, and in dermatitis

herpetiformis, autoantibodies bind to epidermal and tissue transglutaminase. Bullous diseases may present with itching, hive-like welts, and blisters, at times along creases or folds in the skin, depending on the clinical type. The rupture of blisters results in abrasions and crusting. Morbidity and mortality are substantial.<sup>2</sup> These diseases impose a great burden on patients' quality of life with a strong impact on the physical and emotional status.<sup>3-5</sup> A systematic review and meta-analysis on quality of life in pemphigus<sup>6</sup> showed that the greatest impairment was reported in the "symptoms and feelings", and "daily activities" dimen-

sions of the Dermatology Life Quality Index (DLQI), and the highest deterioration was reported in the role-physical, role-emotional, and vitality dimension of SF-36.

The moment of the diagnosis of a life-threatening disease is an emotional time for the patient. At diagnosis, different emotions may arise, such as sadness, despair, fear, confusion, anger, or denial. Post-diagnostic psychological distress has been mainly studied in patients with cancer.<sup>7, 8</sup> In patients with pemphigus vulgaris, it was observed that quality of life was more impaired in the early phase of the disease.<sup>3</sup> However, to our knowledge, specific emotions experienced in that period have not been investigated.

The aim of our study was to investigate the prevalence of different emotions at diagnosis in patients with bullous diseases, in relation to clinical and demographic variables.

### Materials and methods

Adult patients with bullous conditions were recruited through the Italian Association of patients with pemphigus and pemphigoid (ANPPI). Inclusion criteria were: having been diagnosed with a bullous condition by a clinician; age  $\geq 18$  years; signed informed consent. Data were collected by ANPPI through an online questionnaire. Information was collected on sociodemographic data (age, sex, weight, height, education, occupation, marital status) and habits (smoking, alcohol). Clinical information concerned general health status (comorbidities and treatments for other conditions), and characteristics of the bullous condition (phase of the disease, severity, duration, localization). Among several questions concerning psychological issues, it was asked which emotions the patient experienced at the time of the diagnosis: isolation, anger, confusion, sadness, despair, disregard, fear, avoidance, or challenge. Also, the patient reported to whom she/he talked as soon as she/he had the diagnosis: the partner, a relative, a friend, the physician, nobody. It was asked to the patient if she/he considered her/himself as confident, insecure, determined, joyful, introverted, or extroverted.

Categorical data were described as number and percentages. Percentages were compared between subgroups of patients according to sociodemographic and clinical variables using Chi-square Test. P value  $< 0.05$  was considered as significant.

### Results

The study population consisted of 105 patients. Sociodemographic characteristics are described in Table I. Sixty-

TABLE I.—Sociodemographic characteristics of the study population of 105 patients with bullous diseases.

Variable	N.	%
Sex		
Male	38	36.2%
Female	67	63.8%
Age		
18-35 years	5	5.9%
36-44 years	33	38.8%
46-55 years	27	31.8%
56-70 years	9	10.6%
>70 years	10	12.9%
Marital status		
Single	10	9.5%
With partner	78	74.3%
Separated/divorced	12	11.4%
Widow/er	5	4.8%
Educational level		
Primary/secondary	27	25.7%
High school	42	40.0%
University	36	34.3%
Has children		
Yes	77	73.3%
No	28	26.7%
Smoker		
Yes	12	11.4%
No	76	72.4%
Previous	17	16.2%

seven (63.8%) patients were women, and most of them (70.6%) were aged between 36 and 55 years; 74.3% had a partner, 74.3% attended high school or university, 73.3% had children, and 11.4% were smokers. Clinical characteristics of patients are summarized in Table II. Most patients had pemphigus vulgaris, and the second most common condition was pemphigoid. The disease was severe/very severe in 52.4% of patients, and it was active in 12.4% of them. In the other cases it was under pharmacological or medical control or in remission. Age at diagnosis was homogeneously distributed among the different categories, from less than 25 years to 65 years or higher. The most frequent localization at onset was the mouth, and the time from onset to the diagnosis was 2 years or more in 27.6% of cases.

The emotion most frequently experienced at diagnosis (Table III) was confusion (47.6% of patients). Then, more than 30% of patients reported having experienced sadness and fear, 12.5% anger, and 10.5% despair. A lower prevalence was observed for isolation, disregard, avoidance, and challenge. The comparison of the prevalence of the emotions between men and women (Figure 1) revealed that a significantly higher percentage of women than men experienced isolation ( $P=0.038$ ) and despair ( $P=0.043$ ). Also, sadness was close to statistical significance ( $P=0.064$ ).

TABLE II.—Clinical characteristics of the study population of 105 patients with bullous diseases.

Variable	N.	%
<b>Bullous disease</b>		
Pemphigus vulgaris	79	75.2%
Pemphigoid	17	16.2%
Mucous membrane pemphigus	2	1.9%
Pemphigus foliaceus	4	3.8%
Epidermolysis bullosa acquisita	1	1.0%
Pemphigoid and pemphigus	2	1.9%
<b>Disease severity</b>		
Mild	12	11.4%
Moderate	38	36.2%
Severe	40	38.1%
Very severe	15	14.3%
<b>Phase of the disease</b>		
Active	13	12.4%
Pharmacological control	37	35.2%
Medical control, no treatment	20	19.0%
Remission	35	33.3%
<b>Age at onset</b>		
≤25 years	11	10.5%
26-35 years	13	12.4%
36-45 years	27	25.7%
46-55 years	27	25.7%
56-65 years	16	15.2%
>65 years	11	10.5%
<b>Localization at onset (more answers possible)</b>		
Face	22	21.0%
Mouth	74	70.5%
Scalp	45	42.9%
Breast	41	39.0%
Back	47	44.8%
Genitals	24	22.9%
Legs	19	18.1%
<b>Time from onset to diagnosis</b>		
1 month	19	18.1%
3 months	33	31.4%
6 months	16	15.2%
1 year	8	7.6%
≥2 years	29	27.6%
Bullous disease in family	6	5.7%
Other dermatologic disease	39	37.1%

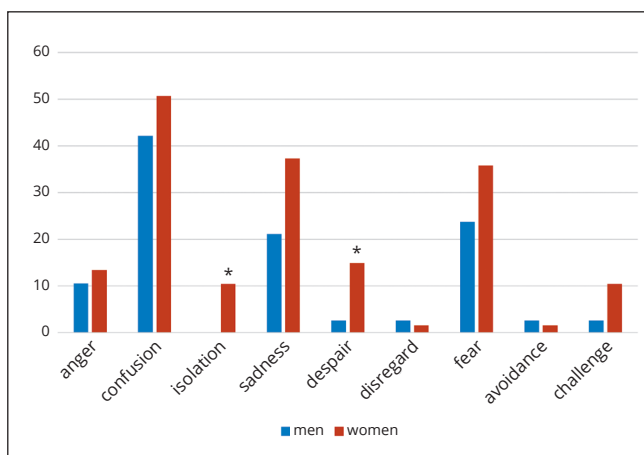


Figure 1.—Prevalence of emotions at diagnosis in men and women with bullous disease. \*P<0.05 (Chi-square Test).

Some differences in the prevalence of emotions (Table IV) were observed for the localization on genitals (more anger, confusion, isolation, fear), having children (more sadness and despair, less avoidance and challenge), and having a relative with a bullous disease (less fear, more challenge). Moreover, the feelings of isolation and fear were more frequent when the lesions were on the breast or the back, and despair when the lesions were on the back. Despair was more frequent in patients who were older at diagnosis. No differences were observed in the frequency of emotions in different categories of age at onset (using both the cutoff

TABLE IV.—Prevalence of “talking with somebody at diagnosis.”

Talking with somebody at diagnosis (more answers possible)	N. (%)
Partner	60 (57.1%)
Relative	25 (23.8%)
Friend	11 (10.5%)
Physician	41 (39.0%)
Nobody	2 (1.9%)

TABLE III.—Reactions to diagnosis in 105 patients with bullous diseases.

Emotions at diagnosis (more answers possible)	N. (%)	Localization on genitals		Has children		Relative with bullous disease	
		Yes	No	Yes	No	Yes	No
Confusion	50 (47.6%)	18 (75.0%)	32 (39.5%)**	36 (46.8%)	14 (50.0%)	2 (33.3%)	48 (48.5%)
Fear	33 (31.4%)	11 (45.8%)	22 (27.2%)*	25 (32.5%)	8 (26.8%)	0 (0.0%)	33 (33.3%)*
Sadness	33 (31.4%)	9 (37.5%)	24 (29.6%)*	28 (36.4%)	5 (17.9%)*	1 (16.7%)	32 (32.3%)
Anger	13 (12.4%)	7 (29.2%)	6 (7.4%)**	10 (13.0%)	3 (10.7%)	1 (16.7%)	12 (12.1%)
Despair	11 (10.5%)	3 (12.5%)	8 (9.9%)*	11 (14.3%)	0 (0.0%)**	0 (0.0%)	11 (11.1%)
Isolation	7 (6.7%)	4 (16.7%)	3 (3.7%)**	5 (6.5%)	2 (7.1%)	0 (0.0%)	7 (7.1%)
Avoidance	2 (1.9%)	0 (0.0%)	2 (2.5%)	0 (0.0%)	2 (7.1%)*	0 (0.0%)	2 (2.0%)
Disregard	2 (1.9%)	0 (0.0%)	2 (2.5%)	2 (2.6%)	0 (0.0%)	0 (0.0%)	2 (2.0%)
Challenge	8 (7.6%)	1 (4.2%)	7 (8.6%)	3 (3.9%)	5 (17.9%)**	2 (33.3%)	6 (6.1%)*

\*P<0.10; \*\*P<0.05, Chi-square Test.

of 35 and 45 years), diagnosis, time from onset to diagnosis ( $\geq 2$  vs.  $< 2$  years), disease severity (severe/very severe versus mild/moderate), and in patients with pemphigus vulgaris compared to pemphigoid (results not shown). When receiving the diagnosis, patients often talked with the partner (57.1%) or with the physician (39.0%), and less frequently with a relative or a friend. Men talked about the diagnosis with their partner more often than women (Figure 2), while women tended to talk more with a relative or the physician. As to personality traits, 23.8% of patients defined themselves as confident, 18.1% insecure, 32.4% determined, 19.0% joyful, 16.2% introverted, and 24.8% extroverted. Emotions experienced by patients with different personality characteristics (Figure 3) were not particularly dissimilar. However, joyful people reported more frequently confusion, extroverted patients reported less frequently fear, and determined people reported less frequently fear and sadness, and more often challenge.

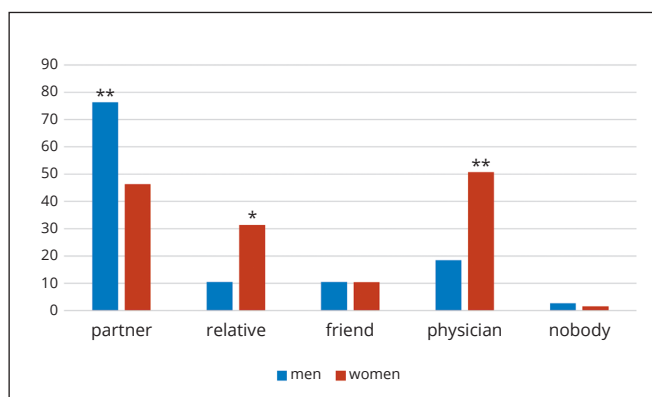


Figure 2.—Prevalence of “talking with somebody at diagnosis” in men and women with bullous disease. \* $P < 0.05$ ; \*\* $P < 0.01$  (Chi-square Test).

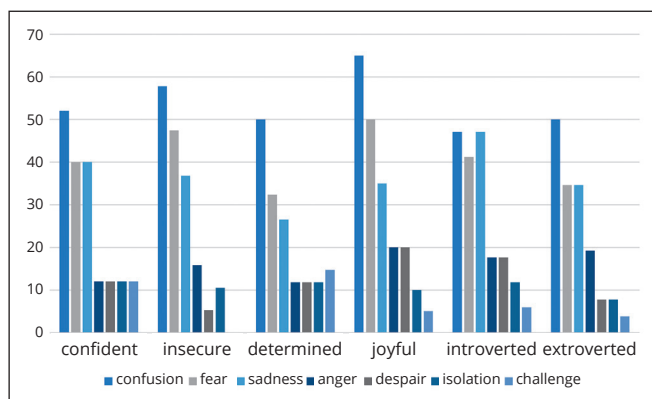


Figure 3.—Prevalence of emotions according to patients' personality characteristics.

## Discussion

In this study we observed that patients with bullous diseases experienced different emotions when receiving the diagnosis of the disease. The emotion that was reported more frequently was confusion, followed by sadness and fear. There is not a unique definition of “emotion”, and this word is often used as a synonym of “feeling.” Emotions precede feelings. They originate in the subcortical regions of the brain, the amygdala, and the ventromedial prefrontal cortices, while feelings originate in the neocortical regions of the brain. Emotions are associated to the body, while feelings to the mind. Paul Ekman defined emotions as “a process, a particular kind of automatic appraisal influenced by our evolutionary and personal past, in which we sense that something important to our welfare is occurring, and a set of psychological changes and emotional behaviors begins to deal with the situation.” Different classifications of emotions have been proposed. According to Paul Ekman,<sup>9</sup> there are six basic emotions: anger, disgust, fear, sadness, surprise, and enjoyment. In the present study, we investigated three of those primary emotions, which could possibly originate at the time of the diagnosis of a severe condition, *i.e.*, as anger, fear, and sadness. According to the classification by Shaver *et al.*,<sup>10</sup> despair and isolation are secondary emotions of sadness. Confusion happens when people face something unknown or difficult to understand. It has been defined as a knowledge emotion,<sup>11</sup> since it appears when something unexpected or believed happens, but at the same time it is fundamental to learning. Challenge arises from a negative experience, however it may be considered a positive emotion, since it represents the possibility to attain a goal.

When receiving a diagnosis of a life-threatening illness or a chronic disease, patients may be flooded with a wide variety of emotions. On one side, the patient may emotionally break down and experience sadness and despair. On the other side, reactions such as anger and challenge may arise, and help cope with the stress caused by the diagnosis. Defense mechanisms can also be put in place, such as removal, denial, and avoidance. The diagnosis of a severe condition may be seen as a type of grief. It is the loss of health and independence, it may result in the loss of work abilities and of a normal social life and may also involve the loss of the usual physical appearance.

Autoimmune bullous disorders are severe, life-threatening conditions, that may strongly impair social life and body image.<sup>12</sup> At the time of diagnosis, despite the explanations of the clinician, the course and the consequences of the disease are still unknown for the patient. He/she has experienced symptoms and quality of life impairment but



may feel confused face to a word that has very likely never heard before. Fear is a common emotion when something unknown appears in one's life. Also, sadness may arise from the feeling that life will not be the same.

To our knowledge, emotions at diagnosis in patients with bullous diseases have not been previously investigated. Feelings and emotions experienced during the course of the disease depend on the daily problems that the disease causes, on the accumulation of stress, frustration and fear towards the future. In a qualitative study,<sup>13</sup> it has been shown that patients with pemphigus experienced distortion in body image, internalized stigma, social isolation, and coping difficulties. However, at the time of the diagnosis, the patient is caught by a whirlwind of emotions.

In the present studies, we observed that some emotions were more frequent in different subgroups of patients. For example, women experienced isolation and despair at diagnosis more often than men. Also, despair was a more frequent reaction in older patients compared to younger ones.

The frequency of emotions, in particular anger, confusion, isolation, and fear, was higher when the disease was localized on genitals compared to other areas. Skin lesions in sensitive areas may result in quality-of-life impairment greater than that of patients with lesions elsewhere on their body, as it is the case for psoriasis.<sup>14</sup>

Emotions were also associated with two important aspects of patients' life, *i.e.*, having children, and having a relative with a bullous condition. Patients may be afraid of not being able to take care of their children due to the disease and may be worried that the child may develop the disease, too. Having a relative with the disease means that the patient already knows the disease, and thus she/he may be less scared and have a more challenging attitude compared to a patient for whom the diagnosis is completely unknown.

Emotions experienced by patients are associated to several factors, such as coping abilities and personality traits. We did not evaluate such aspects however we asked the patients to define themselves using some adjectives, and we observed some differences. For example, people who defined themselves as determined reported less frequently fear and sadness, and more often a challenging attitude. Also, extroverted patients reported less frequently despair and isolation. In previous studies, it has been observed that higher degrees of resilience and extraversion were associated with less psychological distress and loneliness.<sup>15, 16</sup>

### Limitations of the study

A limitation of the study is that patients were at different stages of the disease, and thus recall of emotions at diagno-

sis may be not accurate. However, a strong emotion tied to a significant traumatic event in life is generally remembered with adequate precision also after a long time. Another limitation is that data were collected through an online survey, and thus clinical information was reported by the patients.

### Conclusions

In conclusion, when communicating a diagnosis of a severe condition, such as a bullous disease, it is important to be aware of the emotions of the patient. This is necessary also since emotional states may interfere with processing of information. The clinician should ensure that the patient gets correct information on the disease and that she/he is not overwhelmed with emotions. This can be achieved by active listening, which is at the basis of a good patient-clinician communication.

### References

1. Baum S, Sakka N, Artsi O, Trau H, Barzilai A. Diagnosis and classification of autoimmune blistering diseases. *Autoimmun Rev* 2014;13:482–9.
2. Kridin K, Sagi SZ, Bergman R. Mortality and Cause of Death in Patients with Pemphigus. *Acta Derm Venereol* 2017;97:607–11.
3. Ghodsi SZ, Chams-Davatchi C, Daneshpazhooh M, Valikhani M, Esmaili N. Quality of life and psychological status of patients with pemphigus vulgaris using Dermatology Life Quality Index and General Health Questionnaires. *J Dermatol* 2012;39:141–4.
4. Paradisi A, Sampogna F, Di Pietro C, Cianchini G, Didona B, Ferri R, *et al.* Quality-of-life assessment in patients with pemphigus using a minimum set of evaluation tools. *J Am Acad Dermatol* 2009;60:261–9.
5. Tabolli S, Mozzetta A, Antinone V, Alfani S, Cianchini G, Abeni D. The health impact of pemphigus vulgaris and pemphigus foliaceus assessed using the Medical Outcomes Study 36-item short form health survey questionnaire. *Br J Dermatol* 2008;158:1029–34.
6. Rencz F, Gulácsi L, Tamási B, Kárpáti S, Péntek M, Baji P, *et al.* Health-related quality of life and its determinants in pemphigus: a systematic review and meta-analysis. *Br J Dermatol* 2015;173:1076–80.
7. Beesley VL, Smithers BM, Khosrotehrani K, Khatun M, O'Rourke P, Hughes MC, *et al.* Supportive care needs, anxiety, depression and quality of life amongst newly diagnosed patients with localised invasive cutaneous melanoma in Queensland, Australia. *Psychooncology* 2015;24:763–70.
8. Beesley VL, Hughes MC, Smithers BM, Khosrotehrani K, Malt MK, von Schuckmann LA, *et al.* Anxiety and depression after diagnosis of high-risk primary cutaneous melanoma: a 4-year longitudinal study. *J Cancer Surviv* 2020;14:712–9.
9. Ekman P. Are there basic emotions? *Psychol Rev* 1992;99:550–3.
10. Shaver P, Schwartz J, Kirson D, O'Connor C. Emotion knowledge: further exploration of a prototype approach. *J Pers Soc Psychol* 1987;52:1061–86.
11. Silvia P. Confusion and interest: the role of knowledge emotions in aesthetic experience. *Psychology of Aesthetics, Creativity, and the Arts. Psychol Aesthetics Creativity Arts* 2010;4:75–80.
12. Mazzotti E, Mozzetta A, Antinone V, Alfani S, Cianchini G, Abeni D. Psychological distress and investment in one's appearance in patients with pemphigus. *J Eur Acad Dermatol Venereol* 2011;25:285–9.

13. Okcin F, Ugur O. What Does It Mean To Be Pemphigus Patient? A Qualitative Study. *Clin Nurs Res* 2021;30:790–8.
14. Cengiz FP, Emiroglu N, Wellenhof RH. Dermoscopic and clinical features of pigmented skin lesions of the genital area. *An Bras Dermatol* 2015;90:178–83.
15. Chernova A, Frajo-Apor B, Pardeller S, Tutzer F, Plattner B, Haring C, *et al.* The Mediating Role of Resilience and Extraversion on Psychological Distress and Loneliness Among the General Population of Tyrol, Austria Between the First and the Second Wave of the COVID-19 Pandemic. *Front Psychiatry* 2021;12:766261.
16. Jakobsen IS, Madsen LM, Mau M, Hjemdal O, Friborg O. The relationship between resilience and loneliness elucidated by a Danish version of the resilience scale for adults. *BMC Psychol* 2020;8:131.

---

#### *Conflicts of interest*

The authors certify that there is no conflict of interest with any financial organization regarding the material discussed in the manuscript.

#### *Funding*

This research was financially supported, in part, by the grant “Ricerca Corrente 2022” from the Italian Ministry of Health, Rome, Italy.

#### *Authors' contributions*

Francesca Sampogna, Silvia Battisti, Carola Pulvirenti, Giuseppe Formato, and Damiano Abeni have given substantial contributions to the conception and the design of the study. Chiara Scarpulla, Valentina Battisti, and Federica Cosenza performed data acquisition, analysis and interpretation. Francesca Sampogna, Silvia Battisti, and Damiano Abeni drafted the manuscript, and all the other authors revised it critically. All authors read and approved the final version of the manuscript.

#### *Congresses*

This paper was presented as poster at the Gender Medicine Congress that was held in Padua, Italy on September 16<sup>th</sup>-17<sup>th</sup>, 2022.

#### *Acknowledgements*

The authors thank all the patients who participated in this study.

#### *History*

Manuscript accepted: July 14, 2023. - Manuscript revised: April 13, 2023. - Manuscript received: December 23, 2022.