ORIGINAL ARTICLE: GASTROENTEROLOGY

Positive Orientation and Health-related Quality of Life in Adult Patients Born With Anorectal Malformations

AQ1 *Mariana Fernandes, †Dalia Aminoff, *Cristiano Violani, and *Caterina Grano

ABSTRACT

Objectives: Anorectal malformations (ARMs) are rare congenital colorectal anomalies, which may have a negative impact on health-related quality of life (HRQoL) due to long-lasting consequences, like fecal incontinence. The aim of the present study was to test whether a pervasive mode of appraising and viewing life experiences from a positive stance (ie, positivity) mediates the effect of fecal continence on HRQoL.

Methods: Participants were enrolled from the Italian Association for Anorectal Malformations. Adult patients with ARMs who completed measures of fecal continence (Hirschsprung Disease/Anorectal AQ4 Malformation Quality of Life Questionnaire), positivity (~~SWL, RSES, and LOT~~), and mental/physical HRQoL (SF-36) were included. Data were analyzed using the PROCESS macro for SPSS statistical software (Model 4).

Results: The study included 66 adult patients with ARMs. Mediated regression analyses showed a significant total effect in which patients with higher fecal continence perceived higher physical ($\beta = 0.210$, SE = 0.038, 95% CI [0.133, 0.286]) and mental HRQoL ($\beta = 0.226$, SE = 0.056, 95% CI [0.115, 0.338]) than patients with lower fecal continence. The indirect effects were also significant, indicating that positivity mediated the impact of fecal continence on physical ($\beta = 0.026$, SE = 0.017, 95% CI [0.002, 0.068]), and mental HRQoL ($\beta = 0.146$, SE = 0.058, 95% CI [0.047, 0.275]).

Conclusions: The findings extend previous literature on ARM patients and additional evidence that an optimistic view of oneself, one's life, and one's future contribute to explain the effects of functional impairments on quality of life.

Key Words: anorectal malformations, chronic disease, fecal incontinence, health-related quality of life, positivity

(*JPGN* 2020;70: 00–00)

What Is Known

- Fecal incontinence significantly impairs the physical and mental components of quality of life.
- Positivity is recognized as a basic psychological disposition able to determine better health outcomes.

What Is New

- It is the first study analyzing positivity in patients born with anorectal malformations.
- Positivity partially mediates the relation between fecal continence and physical and mental health-related quality of life.

Anorectal malformations (ARMs) are a complex group of congenital anomalies of the anus and the rectum affecting newborns (1). Despite corrective surgery performed in infancy, many patients born with ARM continue to experience disturbances up to adulthood (eg, fecal incontinence, impaired bowel function, and sexual functioning), with fecal incontinence being the most frequent (2,3). These impairments may have adverse consequences on quality of life (QoL) (2,4–6), although there is evidence that personal and social characteristics (ie, social support, self-efficacy, and coping strategies) play a role in counteracting the effects of fecal incontinence on health-related quality of life (HRQoL) (7–9).

In the last decade, with the growth of theoretical and empirical knowledge in the field of positive psychology (10), increased attention has been given to individual dispositions that may exert a protective role for well-being and individual adjustment. In particular, this literature has focused on positivity, as a basic disposition to view oneself, one's life, and one's future under a positive outlook (11,12) and as a factor~~,~~ which captures the common core of ~~3~~ constructs, namely self-esteem, optimism, and life satisfaction.

Findings have provided evidence that positivity influences several life domains, such as social adjustment, psychological well-being, and health outcomes (13–15). In a sample of cancer patients, Caprara et al (16) reported that higher positive scores were related with a better QoL in terms of less physical and psychological functional impairment both at diagnoses and at 1-year follow-up. Furthermore, recent longitudinal findings have shown the major contribution of positivity to the prediction of health, resiliency, and positive affectivity over an extended length of time in the transition from adolescence to adulthood (13).

To the best of our knowledge, no studies evaluated the role of positivity in determining a better QoL in patients with

Received January 29, 2020; accepted May 17, 2020.

AQ2 From the *Department of Psychology, Sapienza University of Rome, and the †Italian Parents and Patients Organization for Anorectal Malformation (AIMAR), Rome, Italy.

Address correspondence and reprint requests to Caterina Grano, PhD, AQ3 ~~Associate Professor,~~ Department of Psychology, Sapienza University of Rome, Via dei Marsi 78, 00185 Rome, Italy (e-mail: caterina.grano@uniroma1.it).

~~M.F. and C.G. share first authorship.~~

Supplemental digital content is available for this article. Direct URL citations appear in the printed text, and links to the digital files are provided in the HTML text of this article on the journal's Web site (*www.jpgn.org*).

The authors report no conflicts of interest.

DOI: 10.1097/MPG.0000000000002803

gastroenterological diseases. In the context of ARM, some authors, however, highlighted the contribution of one of its components, namely self-esteem, in mitigating the influence of physical symptoms on HRQoL (8,17) and in determining enhanced mental health over time (18). Appraising personal experiences under a positive view may represent a crucial individual strength, which may help to handle or to buffer the effects of ARM impairments on health outcomes. Therefore, the main aim of this study is to explore whether positivity mediates the relation between fecal continence and ~~pysical~~ and mental HRQoL.

METHODS

Participants and Procedure

Adult patients born with ARMs were recruited through the Italian Patients' and Parents' Association for Anorectal Malformations (AIMAR), which is the only ARM patient association in Italy. The patients' members of the association were born with ARM and underwent surgical procedures and follow-ups for these malformations in surgical centers across Italy. Participants received by ordinary mail an envelope including a brief presentation of the research study and the questionnaires. Once they were completed, patients returned the questionnaires using a prepaid envelope. In the present study, all the families with a patient older than16 years with ARM were included. A total of 160 patients received the questionnaire by mail. Of these patients, 71 adult patients born with ARM completed the questionnaires, for a response rate of 44.4%.

The ethical committee of the Department of Psychology, ''Sapienza'' University, approved the study, and only the participants who signed the informed consent were included in the study. In the case of underage patients, one of the parents (usually the mother) signed the informed consent. These same subjects have been previously reported, but with different primary objectives (9,19).

MEASURES

Sociodemographic Information

Each respondent provided sociodemographic and clinical information including age, sex, education, marital status, type of fistula, and associated malformations.

Fecal Continence

Fecal continence was assessed through the 8-item fecal incontinence scale of the Hirschsprung Disease/Anorectal Malformation Quality of Life (HAQL; Italian version) questionnaire (20). The HAQL questionnaire has been validated by Hanneman et al (6) and was adapted to the Italian ARM population with the agreement of the Hanneman group, showing good reliability and concurrent and discriminant validity with other measures (20,21).

An example of an item is ''How frequently do you lose feces before reaching the toilet?'' For each item, participants rated on a 5-point scale (1 = Never; 5 = Always) how frequently the tapped item occurred in the previous 7 days. First, answers to the items of the HAQL were recorded and linearly converted into a 0 to 100 scale (6), with higher scores indicating more fecal continence. The final score was calculated by averaging the items. In the present study, the alpha of the scale was 0.91, which indicated very good reliability.

Positivity

Positive orientation was measured with 3 scales: Life satisfaction, self-esteem, and optimism. These 3 scales were previously validated also in the Italian population (22–24) and have been widely used in both healthy individuals (11,15) and clinical samples (25,26).

Participants' life satisfaction was measured through the ''Life Satisfaction Scale'' (27). For each of the 5-items, participants rated the extent to which they felt generally satisfied with life on a 5-point scale, ranging from 1 (strongly disagree) to 5 (strongly agree). An example item was ''In most ways, my life is close to my ideal'' and ''The conditions of my life are excellent.'' The α coefficient was 0.82, indicating good reliability.

Self-esteem was evaluated through the Rosenberg self-esteem scale (28). The scale included 10-items measured on a 5-point scale (1 = strongly disagree to 5 = strongly agree). Five of the items were positively worded statements and 5 were reversed. An example of an item was ''I feel that I have a number of good qualities.'' The α coefficient was 0.87, indicating good reliability.

Optimism was measured through the ''Life Orientation Test'' (29). The 6-items of the scale assessed subjects' expectations about their future and their general sense of optimism. Respondents provided their ratings by using a 5-point scale ranging from 1 (strongly disagree) to 5 (strongly agree). An example item was ''I always look on the bright side of things.'' The α coefficient was 0.80, indicating good reliability.

The final score of positivity was calculated as a mean of the life satisfaction, self-esteem, and optimism scores.

Health-related Quality of Life

The Italian Short-Form 36 questionnaire was used to evaluate HRQoL (SF-36, v.2) (30). The SF-36 is a measure of health status well-validated (30,31) both in healthy individuals (30,31) and in clinical samples (30,32). It includes 8 domains namely Physical functioning, Role-physical, Bodily pain, Vitality, General health, Social functioning, Role-emotion, and Mental health. For each QoL domain, the item scores were coded, summed, and transformed into a scale from 0 to 100. According to the SF-36 manual, the Physical and the Mental Components were calculated using standard weights, to interpret the summary scales comprehensively (33). The final scores varied from 0 to 100, and higher scores indicated better HRQoL.

Data Analysis

Descriptive analyses, correlations, and analysis of variance were conducted with the statistical program SPSS for Windows version 24.0 (IBM Corp, Armonk, NY). The questionnaires with missing data in critical items were excluded.

To examine whether participants who completed the questionnaire differ from those who did not participate, independent *t* test (~~Student~~ *t*) and χ^2 test were used comparing their sex, age, and geographical area as registered in the AIMAR database.

Considering the data reported in the questionnaires completed, frequencies were calculated for sex, educational level, and marital status. Pearson correlation coefficients were tested to examine the relations between age, educational level, number of surgeries, fecal continence, positive orientation, and physical and mental HRQoL. To analyze differences across sex for fecal continence, positive orientation, and HRQoL (Physical and Mental components), *t* tests were computed. *P* values <0.05 were considered significant.

Subsequently, mediation analyses were conducted using the PROCESS macro for SPSS (Model 4) developed by Hayes (34). Mediation is used to explore the underlying mechanism or process by which one variable influences another variable through a mediator variable (35). Thus, a mediating variable transmits the effect of

an independent variable on a dependent variable (35). Two models were tested, considering as independent variable the fecal continence score and as outcome variables the physical and the mental HRQoL scores, respectively. In both models, the mediation of positivity was analyzed. According to the decomposition of effects approach (36), the indirect effect was calculated using 5000 bootstrap samples for the bootstrap confidence intervals corrected for bias. An indirect effect is considered statistically significant if the confidence intervals established (95%) do not include the value zero (34). The estimated effects reported were unstandardized regression coefficients.

RESULTS

Seventy-one patients completed the questionnaires. In a preliminary analysis, 5 questionnaires had missing data on key items and therefore were discarded. Of the remaining sample (n = 66), 26 were women (39.4%) and 40 were men (60.6%). The mean age was 25.56 (SD = 8.24). Additional characteristics (eg, educational level, marital status) are reported in the supplementary material (Supplemental Table 1, Supplemental Digital Content, *http://links.lww.com/MPG/B858*).

Considering differences between respondents and nonrespondents, no differences in sex ($\chi^2 = 0.948$, $P = 0.33$) and geographical area ($\chi^2 = 3.953$; $P = 0.14$) were observed between respondents and nonrespondents, whereas a significant difference in age was found where the nonrespondents had a lower mean age ($M = 21.50$; SD = 5.03) compared to those who answered the questionnaire ($M = 25.56$; SD = 8.24) (t [110] = −2.975; $P = 0.004$).

The number of surgeries in those who answered the questionnaire ranged from 1 to 16 with a mean of 4.56 (SD = 2.71).

The types of fistulas (using the Krickenbeck classification system) and continence level are reported in Table 1. As can be noticed, the means of fecal continence for some severe types of fistulas (ie, recto bladder neck fistula) are quite high, probably due to the fact that these patients reach continence using irrigations systems.

Frequencies of associated anomalies are reported in Supplemental Table 2 (Supplemental Digital Content, *http://links.lww.com/MPG/B858*) in the supplementary material. The most frequent associated anomalies reported by the patients are urologic and orthopedic anomalies. Considering associated anomalies as a whole, 68.2% (n = 45) of the patients referred to have at least 1 additional congenital anomaly, and 31.8% (n = 21) report not having associated defects. Finally, 68.2% (n = 45) reported more than 1 associated malformation.

In the t tests that analyze whether in the respondents sample there were differences across sex, no significant differences emerged: fecal continence ~~scale~~ (t [64] = 0.194; $P = 0.85$); Physical HRQoL (t [64] = 0.783; $P = 0.44$); Mental HRQoL (t [64] = 1.953; $P = 0.06$); and ~~p~~positivity (t [64] = 1.11; $P = 0.28$).

Before considering mediation regression analyses, correlations among fecal continence, positivity, physical and mental HRQoL with age, educational level, and number of surgeries were also examined and they were all not significant. Fecal continence was positively correlated with positivity and with physical and mental components of HRQoL. Positivity was positively correlated with the physical and mental components of HRQoL. Table 2 presents correlations among the variables.

TABLE 1. Classification of the type of fistulas

	N (%)	Fecal continence Mean (SD)
Imperforate anus	17 (25.8%)	81.25 (12.02)
Perineal fistula	7 (10.6%)	84.29 (19.06)
Rectobulbar urethral fistula	8 (12.1%)	75.39 (24.00)
Recto prostatic urethral fistula	5 (7.6%)	74.38 (25.33)
Recto bladder neck fistula	3 (4.5%)	86.45 (3.61)
Vestibular fistula	5 (7.6%)	66.88 (38.56)
Other type of fistula	5 (7.6%)	84.38 (9.11)
I do not know	5 (7.6%)	86.25 (12.02)
Did not give an answer	11 (16.7%)	79.10 (18.24)

Mediation Analyses

The results of the mediational analyses are presented in Figure 1 and the direct, indirect, and total effects are presented in Table 3. Regarding the mediational model considering as outcome variable the physical HRQoL component, it accounted for 35% of the variance ($R^2 = 0.355$, $F = 17.59$, $P \leq 0.01$). Results showed that Fecal continence significantly predicted positivity ($\beta = 0.122$, SE = 0.004, $P \leq 0.05$). Positivity was not a significant predictor ($\beta = 2.133$, SE = 1.081, $P = 0.053$) of physical HRQoL, and fecal continence significantly predicted physical HRQoL ($\beta = 0.184$, SE = 0.040, $P \leq 0.05$). The total effect of fecal continence on physical HRQoL was significant ($\beta = 0.210$, SE = 0.038, 95% CI [0.133, 0.286]). The indirect effect was significant ($\beta = 0.026$, SE = 0.017, 95% CI [0.002, 0.068]), indicating that positivity mediated the impact of fecal continence on physical HRQoL.

Regarding the mental HRQoL component, the mediation model accounted for 37% of the explained variance ($R^2 = 0.373$, $F = 19.02$, $P \leq 0.01$). Results showed that fecal continence significantly predicted positivity ($\beta = 0.122$, SE = 0.004, $P \leq 0.05$) and that positivity was a significant predictor of mental HRQoL ($\beta = 6.011$, SE = 0.053, $P \leq 0.05$). The effect of fecal continence on mental HRQoL was significant ($\beta = 0.153$, SE = 1.437, $P \leq 0.01$). The total effect of fecal continence on mental HRQoL was significant ($\beta = 0.226$, SE = 0.056, 95% CI [0.115, 0.338]). The indirect effect was also significant ($\beta = 0.146$, SE = 0.058, 95% CI [0.047, 0.275]), indicating that positivity mediated the impact of fecal continence on mental HRQoL. The standardized parameter estimates of the tested model are reported in Table 3.

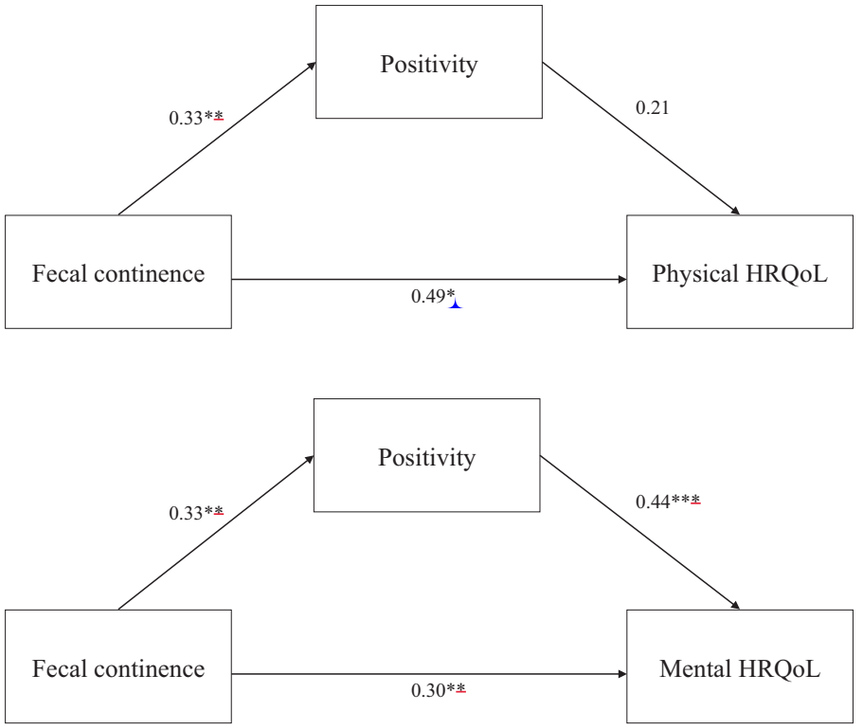
DISCUSSION

Stemming from the positive psychology perspective, the purpose of this study was to estimate the impact of fecal continence on the physical and mental components of HRQoL, evaluating whether positivity mediated these relations.

Our findings provide evidence of a positive and direct path of fecal continence on both HRQoL components. More specifically, fecal continence is related to lower role restrains, enhanced physical functioning, fewer limitations in daily activities (physical HRQoL), and to greater energy and psychological well-being (mental HRQoL). This is consistent with previous studies conducted in patients born with ARM, indicating that fecal incontinence had a direct impact on the different QoL domains (3,5,20).

Considering the mediational role of positivity, and extending previous literature on ARM patients in which only 1 component of positivity was examined (ie, self-esteem) (8,17), our findings support, in addition, that an optimistic view of oneself, one's life, and one's future, partially influence the effect of fecal continence on both physical and mental HRQoL components. It seems that when dealing with impairments or limitations due to a disease, having a more general positive attitude enables patients to cope better with them, leading to enhanced physical and mental health. Indeed, it has

CE: ; JPGN-20-091; Total nos of Pages: 7;
JPGN-20-091



Note: Standardized regression coefficients are reported.

FIGURE 1. Path coefficients in the tested model. HRQoL = health-related quality of life.

been suggested that patients with higher positivity seem to be more prone to use more adaptive cognitive strategies and to benefit from available supports (16). Considering that previous findings have shown that ARM patients using maladaptive coping strategies reported also worse mental health (37), focusing on positivity as a way to use more adaptive coping strategies to deal with disease-related stressors may be beneficial.

The above findings are also in line with the results of previous studies conducted in the context of other diseases, in which positive orientation was associated with better health outcomes and adjustment (16), suggesting that positivity exerted its protective buffering role by attenuating negative thoughts and feelings of hopelessness.

Regarding the practical implications, interventions targeting positivity may be useful to help patients to value better their resources and to cope more effectively with the disease impairments. In fact, despite positivity is considered as a stable disposition, it can be trained through mastery experiences aimed at enhancing its components (ie, self-esteem, life satisfaction, and optimism) (12). Indeed, skills-based psychological interventions to improve self-esteem have been shown to be effective in increasing self-esteem levels and in decreasing psychological distress in patients with other diseases (38).

In our sample, educational level was not associated with HRQoL outcomes. Although in previous studies on ARM conducted with other samples, educational level was included as a measure (39–41), only 1 study considered the direct association with HRQoL (17) and it did not find a significant association, differently from what is reported in other chronic diseases where the educational background has a stronger impact (42–44).

Finally, the present study has some limitations, which need to be acknowledged. First, the study is cross-sectional; thus, causal relations cannot be inferred. Longitudinal studies are needed to examine the causal direction of the observed paths and how the relations between fecal continence, positivity, and HRQoL may change along time. Second, both components of HRQoL were measured through a generic QoL instrument, which does not take into account specific aspects of QoL-related to fecal incontinence, like embarrassment or shame. Third, the participants enrolled in the study were only the members of the AIMAR association and this may limit the representativeness of the sample. In fact, patients with less severe forms of ARM may be underrepresented as they may have not felt in need to ask support from the association. Therefore, the present findings should not be generalized to all the population of ARM patients. Fourth, the sample size was limited. It, however, has to be noted that ARMs are rare malformations and that our sample includes nearly 45% of the whole adult patient members of

TABLE 2. Correlations among fecal continence, positive orientation, physical and mental health-related quality of life

	1	2	3	4	5	6
1. Age	–					
2. Educational level	0.12	–				
3. Number of surgeries	−0.01	0.06	–			
4. Fecal continence	−0.22	−0.10	−0.07	–		
5. Positivity	−0.03	−0.11	0.02	0.35‡	–	
6. Physical HRQoL	−0.12	0.09	−0.10	0.56‡	0.37‡	–
7. Mental HRQoL	−0.07	−0.01	0.03	0.45‡	0.54‡	0.34‡

HRQoL = health-related quality of life.
*$P < 0.01$.
†$P < 0.001$.

TABLE 3. Standardized path coefficients for mediated effects on physical and mental health-related quality of life

		Direct effect		Indirect effect		Total effect	
Paths	Mediator	Std. estimation	95% CI low/high	Std. estimation	95% CI low/high	Std. estimation	95% CI low/high
Fecal continence → physical HRQoL	Positivity	0.184†	0.104/0.263	0.026‡	0.002/0.007	0.210†	0.133/0.286
Fecal continence → mental HRQoL	Positivity	0.153‡	0.047/0.258	0.146‡	0.047/0.275	0.226†	0.115/0.338

HRQoL = health-related quality of life.
*$P < 0.05$.
†$P < 0.01$.‡The indirect effect is significant as the confidence interval do not include the value zero.

the Italian Association, which is the only ARM association in the country. Future studies may help to evaluate how fecal continence and positivity interact in influencing the QoL also in patients belonging to different age groups, as previous studies showed that while fecal continence may improve as patients grow up, the QoL in the psychosocial-related areas may decrease (45,46). Another limitation of the present study was the lack of access to the medical records of the patients born with ARM; therefore, the information regarding the type of fistula and associated defects was only self-reported. Finally, the present study did not consider the influence of patients' social environment, which may have a relevant influence on QoL, neither assess the possible effect of negative traits (eg, depressiveness, social anxiety), which may overlap with positive orientation. Future studies may explore the concomitant role of other significant personality and psychosocial variables (eg, social support, hostility traits) when assessing the protective role of positivity.

CONCLUSIONS

To our knowledge, the present study is the first that explores the role of positivity in adult patients with a rare gastroenterological disease. Our findings, besides supporting previous findings (5,17) on the negative effects of fecal incontinence on HRQoL, broaden previous knowledge, highlighting the protective role of positivity in determining better physical and mental health outcomes. In line with these findings, it may be particularly useful to appraise ARM patients' personal resources to combine medical treatments with effective psychological interventions. Indeed, the early identification of patients' positivity levels may be beneficial to plan specific interventions to support patients with lower positive orientation. Although these conclusions are in line with the literature and with other studies that emphasize the mediational buffering role of positive orientations, the causality of the investigated paths is not demonstrated and future longitudinal studies are needed to better explore the causal relations between fecal continence, positivity, and HRQoL.

Acknowledgments: The authors of the present article and AIMAR Association wish to thank all the members of the AIMAR Association who participated in this study.

REFERENCES

1. Murphy F, Puri P, Hutson JM, et al. Incidence and frequency of different types, and classification of anorectal malformations. In: Hohlschneider AM, Hustson JM, eds. *Anorectal Malformations in Children.* Berlin, Heidelberg: Springer; 2006:163–84.
2. Rintala R, Pakarinen MP. Outcome of anorectal malformations and Hirschsprung's disease beyond childhood. *Semin Pediatr Surg* 2010;19:160–7.
3. Rintala R, Mildh L, Lindahl H. Fecal continence and quality of life for adult patients with an operated high or intermediate anorectal malformation. *J Pediatr Surg* 1994;29:777–80.
4. Athanasakos EP, Kemal KI, Malliwal RS, et al. Clinical and psychosocial functioning in adolescents and young adults with anorectal malformations and chronic idiopathic constipation. *Br J Surg* 2013;100:832–9.
5. Davies MC, Liao L-M, Wilcox DT, et al. Anorectal malformations: what happens in adulthood? *BJU Int* 2010;106:398–404.
6. Hanneman MJG, Sprangers MAG, De Mik EL, et al. Quality of life in patients with anorectal malformation or Hirschsprung's disease. *Dis Colon Rectum* 2001;44:1650–60.
7. Hartman EE, Oort FJ, Aronson DC, et al. Quality of life and disease-specific functioning of patients with anorectal malformations or Hirschsprung's disease: a review. *Arch Dis Child* 2011;96:398–406.
8. Hartman EE, Oort FJ, Aronson DC, et al. Explaining change in quality of life of children and adolescents with anorectal malformations or Hirschsprung disease. *Pediatrics* 2007;119:e374–83.
9. Grano C, Fernandes M, Bucci S, et al. Self-efficacy beliefs, faecal incontinence and health-related quality of life in patients born with anorectal malformations. *Color Dis* 2018;20:711–8.
10. Seligman ME, Csikszentmihalyi M. Positive psychology. An introduction. *Am Psychol* 2000;55:5–14.
11. Caprara GV, Fagnani C, Alessandri G, et al. Human optimal functioning: the genetics of positive orientation towards self, life, and the future. *Behav Genet* 2009;39:277–84.
12. Caprara GV, Steca P, Alessandri G, et al. Positive orientation: explorations on what is common to life satisfaction, self-esteem, and optimism. *Epidemiol Psichiatr Soc* 2010;19:63–71.
13. Alessandri G, Caprara GV, Tisak J. A unified latent curve, latent state-trait analysis of the developmental trajectories and correlates of positive orientation. *Multivariate Behav Res* 2012;47:341–68.
14. Fagnani C, Medda E, Stazi MA, et al. Investigation of age and gender effects on positive orientation in Italian twins. *Int J Psychol* 2014;49:453–61.
15. Alessandri G, Caprara GV, Tisak J. The unique contribution of positive orientation to optimal functioning: further explorations. *Eur Psychol* 2012;17:44–54.
16. Caprara GV, Castellani V, Alessandri G, et al. Being positive despite illness: the contribution of positivity to the quality of life of cancer patients quality of life of cancer patients. *Psychol Health* 2016;31:524–34.
17. Hartman EE, Oort FJ, Aronson DC, et al. Critical factors affecting quality of life of adult patients with anorectal malformations or Hirschsprung's disease. *Am J Gastroenterol* 2004;99:907–13.
18. Hartman EE, Oort FJ, Visser MR, et al. Explaining change over time in quality of life of adult patients with anorectal malformations or Hirschsprung's disease. *Dis Colon Rectum* 2006;49:96–103.
19. Grano C, Bucci S, Aminoff D, et al. Feelings of depression in people with arm: the role of critical incidents and perceived difficulties in close and sexual relationships. *Pediatr Surg Int* 2014;30:823–8.
20. Grano C, Aminoff D, Lucidi F, et al. Long-term disease-specific quality of life in adult anorectal malformation patients. *J Pediatr Surg* 2011;46:691–8.

21. Grano C, Aminoff D, Lucidi F, et al. Disease-specific quality of life in children and adults with anorectal malformations. *Pediatr Surg Int* 2010;26:151–5.
22. Prezza M, Trombaccia FR, Armento L. La scala dell'autostima di Rosenberg: traduzione e validazione Italiana [The Rosenberg Self-Esteem Scale: Italian translation and validation]. *Giunti Organ Spec* 1997;223:35–44.
23. Giannini M, Schuldberg D, Di Fabio A, et al. Misurare l'ottimismo: proprietà psicometriche della versione Italiana del Life Orientation Test—Revised (LOT-R). [Measuring optimism: psychometric properties of the Italian version of the Life Orientation Test—Revised (LOT-R).]. *Couns G Ital di Ric e Appl* 2008;1:73–84.
24. di Fabio A, Palazzeschi L. The Satisfaction With Life Scale (SWLS): un contributo alla validazione italiana con lavoratori adulti. [The Satisfaction With Life Scale (SWLS): a contribution to Italian validation with adult workers.]. *Couns G Ital di Ric e Appl* 2012;5:207–15.
25. ~~Caprara GV, Barbaranelli C, Borgogni L, et al. The ''big five questionnaire'': a new questionnaire to assess the five factor model. *Pers Individ Dif* 1993;15:281–8.~~
26. Curbow B, Somerfield M. Use of the Rosenberg self-esteem scale with adult cancer patients. *J Psychosoc Oncol* 1991;9:113–31.
27. Diener E, Emmons Ra, Larsen RJ, et al. The satisfaction with life scale. *J Pers Assess* 1985;49:71–5.
28. Rosenberg M. Society and the Adolescent Self-image. Princeton, NJ: Princeton University Press; 1965.
29. Scheier M, Carver C. Optimism, coping, and health: assessment and implications of generalized outcome expectancies. *Heal Psychol* 1985;4:219–47.
30. Apolone G, Mosconi P. The Italian SF-36 health survey. *J Clin Epidemiol* 1998;51:1025–36.
31. Brazier JE, Harper R, Jones NM, et al. Validating the SF-36 health survey questionnaire: new outcome measure for primary care. *BMJ* 1992;305:160–4.
32. Kosinski M, Keller SD, Ware JE, et al. The SF-36 Health Survey as a generic outcome measure in clinical trials of patients with osteoarthritis and rheumatoid arthritis: relative validity of scales in relation to clinical measures of arthritis severity. *Med Care* 1999;37:MS23–39.
33. Ware JE, Kosinski M, Keller SD. *SF-36 Physical and Mental Health Summary Scales: A User's Manual. Boston, MA: Health Institute, New England Medical Center; 1994*.
34. Hayes AF. *Introduction to Mediation, Moderation, and Conditional Process Analysis: A Regression-based Approach.* 7th ed. New York, NY: The Guilford Press; 2017.
35. MacKinnon DP, Fairchild AJ, Fritz MS. Mediation analysis. *Annu Rev Psychol* 2007;58:593–614.
36. Alwin DF, Hauser RM. The Decomposition of Effects in Path Analysis. *Am Sociol Rev* 1975;40:37–47.
37. Grano C, Fernandes M, Aminoff D, et al. The role of coping strategies on health-related quality of life in adults with anorectal malformations. *Pediatr Surg Int* 2016;32:759–65.
38. Chadwick PM, Smyth A, Liao L-M. Improving self-esteem in women diagnosed with Turner syndrome: results of a pilot intervention. *J Pediatr Adolesc Gynecol* 2014;27:129–32.
39. van der Bent A, Duggan EM, Fishman LN, et al. Reality check: what happens when patients with anorectal malformations grow up? A pilot study of medical care transition from the adult patient perspective. *J Pediatr Surg* 2018;53:1722–6.
40. Drissi F, Meurette G, Baayen C, et al. Long-term outcome of Hirschsprung disease. *Dis Colon Rectum* 2019;62:727–32.
41. Poley MJ, Stolk E, Tibboel D, et al. Short term and long term health related quality of life after congenital anorectal malformations and congenital diaphragmatic hernia. *Arch Dis Child* 2004;89:836–41.
42. Juan J, Estiarte R, Colomé E, et al. Burden of illness of Crohn's disease in Spain. *Dig Liver Dis* 2003;35:853–61.
43. Iglesias M, Vázquez I, Barreiro de Acosta M, et al. Health related quality of life in patients with Cohn's disease in remission. *Rev Española Enfermedades Dig* 2010;102:624–9.
44. Casellas F, López-Vivancos J, Casado A, et al. Factors affecting health related quality of life of patients with inflammatory bowel disease. *Qual Life Res* 2002;11:775–81.
45. Giuliani S, Grano C, Aminoff D, et al. Transition of care in patients with anorectal malformations: consensus by the ARM-net consortium. *J Pediatr Surg* 2017;52:1866–72.
46. Grano C, Bucci S, Aminoff D, et al. Transition from childhood to adolescence: quality of life changes 6 years later in patients born with anorectal malformations. *Pediatr Surg Int* 2015;31:735–40.

MPG

Manuscript No. JPGN-20-091

Journal of Pediatric Gastroenterology and Nutrition

Dear Author,

During the preparation of your manuscript for typesetting, some queries have arisen. These are listed below. Please check your typeset proof carefully and mark any corrections in the margin as neatly as possible or compile them as a separate list. This form should then be returned with your marked proof/list of corrections to the Production Editor.

QUERIES: to be answered by AUTHOR

QUERY NO.	QUERY DETAILS	RESPONSE
<AQ1>	Please confirm whether surnames/family names (red) have been identified correctly in the author byline.	AQ1: The surnames/family names of the authors are correct.
<AQ2>	Affiliations have been set as per style. Please check for accuracy of information.	AQ2: The affiliation information has been changed
<AQ3>	Please check and confirm the corresponding author's name and relevant details for correctness.	AQ3: The corresponding author's name were correct, but the details has been changed.
<AQ4>	Please spell out the acronyms SWL, RSES, and LOT.	AQ4:The acronyms were deleted in the text, and replaced with the scales' names: "(Life Satisfaction Scale, Rosenberg Self-esteem Scale, and Life Orientation Test)"
<AQ5>	Please check the terms "fecal continence scale" and "fecal continence" for correctness.	AQ5: "Fecal continence" is the correct term.
<AQ6>	References have been updated using PubMed. Please check for correctness of information.	AQ6: All the references are correct, with the exception of the reference number 25.
<AQ7>	Please link footnote "$*$" Table 2.	AQ7: In Table 2, all values are P<0.01, therefore the footnote p<0.001 must be deleted.
<AQ8>	Please link all the foonotes in Table 3.	AQ8: In Table 3, only one value have P<0.05. All footnotes are linked now.