Author's response to reviews

Title: Leiomyomatosis peritonealis disseminata in association with multiple congenital malformations: a new feature of Currarino syndrome?

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Version: 5 Date: 7 March 2006

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Answer to referees

Answer to Referee Sonu Nigam

Page 5, para 2.”Her past history was characterized by several congenital malformations mostly of whom had required surgical treatment.” Should be “…. Most of which had required …..”

The text has been modified according to the referee’s suggestion (Abstract: case presentation and page 4, paragraph 2).

Page 5, para 3 “ In the neonatal age, she had an anoplasty because an imperforate anus with an ectopic vulvar anal orifice.” Should be “ anoplasty for……”

The text has been modified according to the referee’s suggestion (page 4, paragraph 3).

Page 5: No mention of macroscopic appearance of Uterus/hysterectomy.

The text has been modified according to the referee’s suggestion (page 4, paragraph 9 and page 5, paragraphs 2-3).
What is the ER, PR status of the lesion?

**Immunohistochemical evaluation for ER and PR has been performed by C.M.**

This is now clearly stated in the text (page 5; paragraph 4; Figure 4).

Page 8: “In elder study Mathews…..”. Does it mean “another study”?

**The text has been modified according to the referee’s suggestion.**

Page 9: “Beckers hypnotised ……” Is it “hypothesized”?

**The text has been modified according to the referee’s suggestion (Page 9, paragraph 1).**

Page 9: Para3 “If intestinal symptoms…preminent” should be “prominent”.

**The text has been modified according to the referee’s suggestion (Page 9, paragraph 4).**

Page 10: Since the diagnosis of Currarino’s syndrome was made on clinical observations alone, a cytogenetic analysis would have been confirmatory

**Cytogenetic analysis has been performed by Elisa Merello and Valeria Capra at the Gaslini Hospital of Genova, the only laboratory in Italy able to perform such**
analysis. The results (Page 5, paragraph 6; Page 6) and relative discussion (Page 10, paragraph 7; page 11, paragraphs 2-3) have been added in the text.

Answer to Referee Huseyin Ilhan

1. The underlying mechanism of LPD development is not known, but involvement of estrogens as etiologic factors has been postulated. LPD is an acquired disease. For this reason, in title of manuscript, “Leiomyomatosis peritonealis disseminata in association with Currarino syndrome” is more appropriate than “………: a new feature of Currarino syndrome?” The association of Currarino syndrome with LPD must be emphasized instead of a new feature of Currarino syndrome.

The title has been modified according to the referee’s suggestion.

2. This case is incomplete Currarino syndrome: hemisacrum and only one of the other malformations, i.e., probably, anorectal malformation with rectovestibular fistula.

The text has been modified according to the referee’s observation and the recent classification of Currarino syndrome (Page 10, paragraph 4).

3. “ectopic vulvar anal orifice” or “vulvar anus” are not accurate terms; both must be corrected according to Péna classification, i.e., probably, anorectal malformation with rectovestibular fistula.
The text has been modified according to the referee’s suggestion (page 4, paragraph 3).

3. “pielonephrite” must be “pyelonephritis”

The text has been modified according to the referee’s suggestion (page 4, paragraph 5).

4. The number for references must be given arranged in order, for example, [4, 6], [6, 9], [7, 11] or must be written at the end of related sentence.

The references have been modified according to the referee’s suggestion.

5. The conclusion is containing too classical knowledge; the association of Currarino syndrome and LPD must be discussed in detail.

The discussion has been mostly rewritten and the conclusions have been modified.

**Answer to Referee Francisco F Nogales**

The authors fail to prove this assumption because:

1. They report a multinodular pelvic mass but certainly not a case of LPD which consists in the presence of many independent nodules throughout the peritoneum. As
to the origin of mass, they do not state location or relationships. The presence or involvement of the uterus is not even mentioned.

The masses have been clearly described in the revised manuscript (Page 5, paragraphs 1-3). The histological diagnosis of LPD has been also confirmed by Professor Jaime Prat, Director of the Department of Pathology, Hospital de la Santa Creu i Sant Pau, Autonomous University of Barcelona, Spain.

2 The association of the mass with the malformation might be coincidental unless otherwise proven.

The manuscript has been completely revised and this issue has been addressed in the Discussion section.