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*Article*

# Syndromic vs. Non-Syndromic Jejunojejunal Intussusception in Adolescents: Clinical and Pathological Insights from a Tertiary Care Centre in Maharashtra


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*Abstract*: ***Introduction***: Jejunojejunal intussusception is a rare anatomical subtype of bowel telescoping in adolescents. Unlike idiopathic intussusception seen in infants, adolescent cases often involve a pathological lead point. Distinguishing between syndromic and non-syndromic etiologies is essential, given their differing clinical implications and management strategies. ***Objective***: To compare clinical presentation, diagnostic features, pathological findings, and outcomes between syndromic and non-syndromic jejunojejunal intussusception in adolescents. ***Methodology***: This retrospective observational comparative study was conducted over five years at a tertiary care hospital. All adolescents aged 10–19 years who underwent surgical treatment for jejunojejunal intussusception were included. Data were extracted from operative, histopathological, and genetic records. Syndromic cases were identified by histological or genetic evidence of polyposis syndromes or systemic features. Statistical analysis was performed using SPSS v25. ***Results***: Of 28 adolescents with jejunojejunal intussusception, 17 (60.7%) were non-syndromic and 11 (39.3%) syndromic. Abdominal pain was universal, while vomiting occurred in 75%. Family history and multiple polyps were confined to syndromic patients ($p < 0.0001$), and recurrence was significantly higher in this group (63.6% vs. 0%, $p = 0.0050$). ***Conclusion***: Syndromic cases are distinguished by family history, multiple polyps, and greater recurrence risk. Accurate classification with histopathology and genetic testing, coupled with structured surveillance, is critical for optimizing long-term outcomes.



*Keywords*: Adolescents, Jejunojejunal intussusception, Polyposis syndrome, Syndromic, Non-syndromic.


## BACKGROUND

Intussusception is a gastrointestinal condition in which one segment of the bowel telescopes into an adjacent segment, leading to bowel obstruction and potentially ischemia if untreated. It is the most common cause of intestinal obstruction in infants and toddlers, but its incidence decreases markedly with age, making adolescent presentations rare. In children under two years, intussusception is typically idiopathic, whereas in older children and adolescents, a pathological lead point such as a polyp, Meckel's diverticulum, or tumor is more often implicated. [1]

Jejunojejunal intussusception represents a particularly uncommon anatomical subtype. Most pediatric cases involve ileocolic segments, with jejunojejunal forms reported only sporadically in the literature, often associated with a definable

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structural lesion. [2] Because of this rarity, recognition may be delayed, and the diagnosis frequently relies on high-resolution imaging or intraoperative findings.

A key distinction in adolescent intussusception involves whether the condition is syndromic or non-syndromic. Syndromic cases typically occur in the context of genetic polyposis disorders such as Peutz–Jeghers syndrome (PJS), characterized by hamartomatous gastrointestinal polyps and mucocutaneous pigmentation. [3] Juvenile Polyposis Syndrome (JPS) presents with multiple hamartomatous polyps distributed throughout the gastrointestinal tract. [4] In contrast, non-syndromic cases are often characterized by solitary polyps or other isolated lead points without systemic features or family history. [1]

Given the prognostic and management implications—syndromic patients often requiring lifelong surveillance, genetic counseling, and family screening—accurately distinguishing between these two categories is crucial. The present study aims to compare syndromic and non-syndromic adolescent jejunojejunal intussusception in terms of clinical presentation, diagnostic findings, pathological characteristics, and outcomes, thereby aiding in timely recognition and tailored management strategies.

**Objective**
To compare the clinical presentation, diagnostic characteristics, pathological findings, and outcomes of syndromic versus non-syndromic jejunojejunal intussusception in adolescents.

## METHODOLOGY
This retrospective observational comparative study was conducted at a tertiary care teaching hospital in [insert location if needed], serving as the study area. The study was carried out over a five-year period, from [start year] to [end year], and included all adolescent patients aged 10–19 years who underwent surgical management for jejunojejunal intussusception, constituting the study population. Each individual patient formed a single study unit. Inclusion criteria comprised adolescents aged 10–19 years with surgically confirmed jejunojejunal intussusception, complete operative and histopathology records, a minimum of three months postoperative follow-up, and sufficient clinical, histological, or genetic data to allow classification as syndromic or non-syndromic. Patients were excluded if intussusception involved other bowel segments, if records were incomplete, if follow-up was less than three months, or if syndromic classification could not be determined. A total enumeration sampling method was employed, with the sampling frame consisting of hospital surgical logbooks, operative notes, and pathology records. All eligible cases during the study period were included, making up the final sample size of eight patients. Data were collected using a structured proforma as the study tool, capturing demographic details, clinical presentation, imaging findings, operative and pathological findings, genetic test results, and postoperative outcomes. Data collection was conducted retrospectively from institutional archives. Ethical clearance was obtained from the Institutional Ethics Committee, and all patient data were anonymized to maintain confidentiality. Operational definitions included classifying cases as syndromic if associated with histopathological or genetic evidence of a polyposis syndrome or systemic features, and non-syndromic when associated with isolated pathology without systemic or familial indicators. Data were analyzed using SPSS version 25.0. Continuous variables were analyzed using the Student's t-test or Mann–Whitney U test based on normality, while categorical variables were assessed using Chi-square or Fisher's exact test, with statistical significance set at $p < 0.05$.

## RESULTS
A total of 28 adolescent patients diagnosed with jejunojejunal intussusception and managed surgically over the five-year study period were included in the analysis. Of these, 17 (60.7%) were classified as non-syndromic and 11 (39.3%) as syndromic cases based on clinical, histopathological, and genetic criteria. The results are presented in the following tables and figures, outlining the demographic profile, clinical presentation, diagnostic and pathological findings, surgical outcomes, and statistical comparison between syndromic and non-syndromic groups. Table 1 shows the demographic profile of the study participants. Out of the total 28 patients, the majority (17; 60.7%) were in the 10–14 years age group, while 11 (39.3%) were in the 15–19 years group. Females constituted 18 (64.3%) of the participants, and males 10 (35.7%). Figure 1 illustrates the distribution of participants based on syndromic status, with 17 (60.7%) being non-syndromic and 11 (39.3%) syndromic cases. Figure 2 shows the distribution of study participants based on clinical presentation. All patients (28; 100%) presented with abdominal pain. Vomiting was reported in 21 (75.0%) patients, while bleeding per rectum was observed in 10 (35.7%). A positive family history was documented in 11 (39.3%) patients, and systemic features were also present in 11 (39.3%).

Table 2 presents the distribution of participants based on investigations and pathological findings. A lead point was detected on imaging in 17 (60.7%) patients. The location of intussusception was equally divided between proximal jejunum (14; 50.0%) and mid jejunum (14; 50.0%). Multiple polyps were found in 11 (39.3%) patients. Histopathology revealed juvenile polyps in 17 (60.7%), hamartomatous polyps suggestive of Peutz–Jeghers syndrome in 7 (25.0%), and inflammatory polyps in 4 (14.3%). Genetic testing was performed in 11 (39.3%) patients, of whom 7 (25.0%) had a positive result. Table 3

 

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outlines surgical outcomes. Segmental resection with anastomosis was the most common procedure performed (25; 89.3%), while 3 (10.7%) underwent polypectomy. Postoperative complications occurred in 3 (10.7%) patients. Recurrence was observed in 7 (25.0%) patients. Follow-up duration was ≥ 12 months in 17 (60.7%) cases and < 12 months in 11 (39.3%).

**Table 4** compares selected variables between syndromic and non-syndromic cases. The sex distribution was similar between the two groups, with females comprising 63.6% of syndromic and 64.7% of non-syndromic patients ($p = 1.0000$). Vomiting was observed in all syndromic patients (100.0%) compared to 58.8% of non-syndromic patients, showing a statistically significant difference ($p = 0.0300$). Family history was positive in all syndromic patients (100.0%) and absent in the non-syndromic group (0.0%), which was highly significant ($p < 0.0001$). Similarly, multiple polyps were found in all syndromic patients (100.0%) and in none of the non-syndromic cases, also demonstrating strong statistical significance ($p < 0.0001$). Recurrence was documented in 63.6% of syndromic patients and absent in the non-syndromic group, representing a statistically significant difference ($p = 0.0050$).

**Table 1: Distribution of study participants based on demographic profile (n = 28)**

| Variable | Category | Frequency (n) | Percentage (%) |
|---|---|---|---|
| **Age Group (years)** | 10–14 | 17 | 60.70% |
| | 15–19 | 11 | 39.30% |
| **Sex** | Female | 18 | 64.30% |
| | Male | 10 | 35.70% |

**Figure 1: Distribution of study participants based on syndromic status**

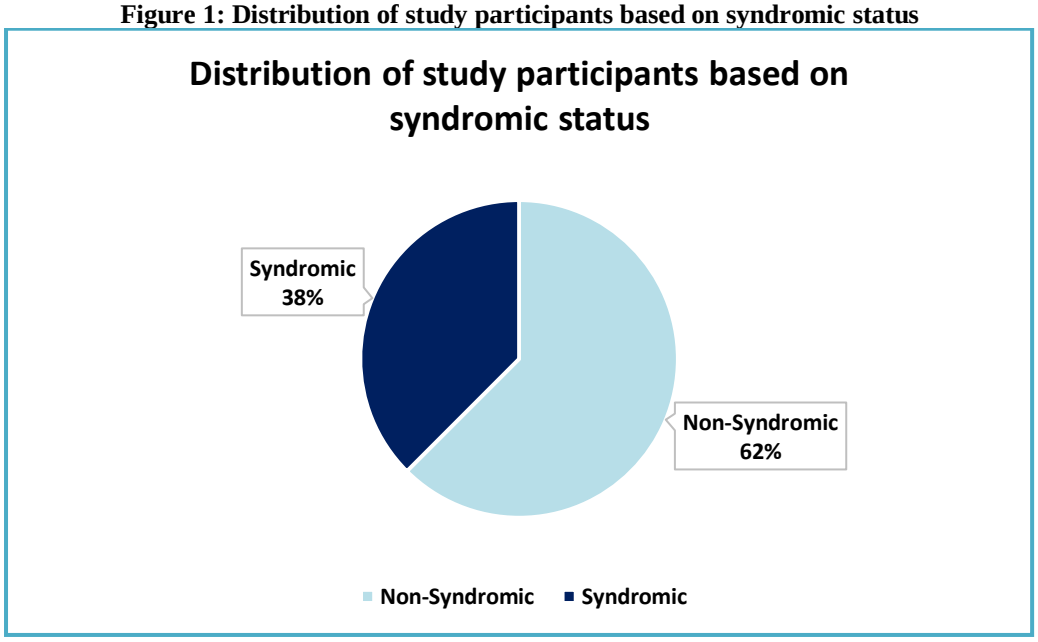


**Figure 2: Distribution of study participants based on clinical presentation**

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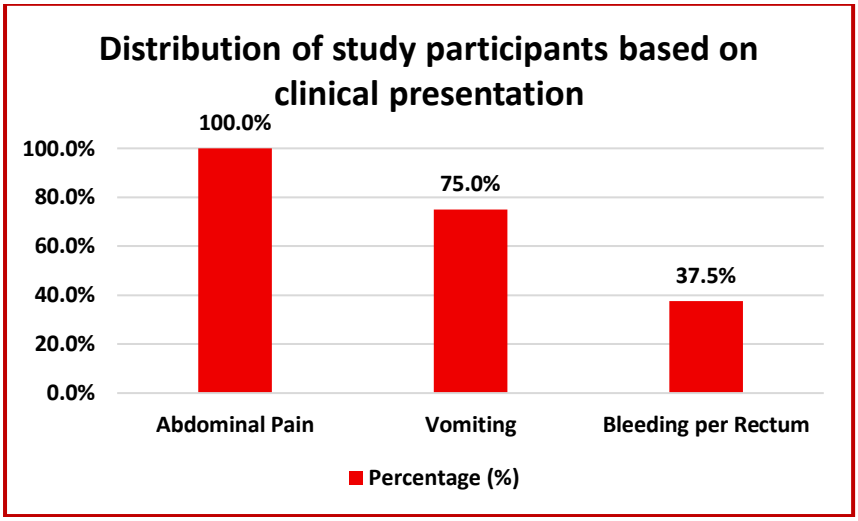


**Table 2: Distribution of study participants based on investigations and pathological findings (n = 28)**

| Investigations and findings | Category | Frequency (n) | Percentage (%) |
|---|---|---|---|
| **Lead Point Detected on Imaging** | Yes | 17 | 60.70% |
| | No | 11 | 39.30% |
| **Location of Intussusception** | Proximal Jejunum | 14 | 50.00% |
| | Mid Jejunum | 14 | 50.00% |
| **Multiple Polyps** | Yes | 11 | 39.30% |
| | No | 17 | 60.70% |
| **Histopathology Diagnosis** | Juvenile Polyp | 17 | 60.70% |
| | Hamartomatous Polyp (PJS) | 7 | 25.00% |
| | Inflammatory Polyp | 4 | 14.30% |
| **Genetic Test Done** | Yes | 11 | 39.30% |
| | No | 17 | 60.70% |
| **Positive Genetic Result** | Yes | 7 | 25.00% |
| | No | 21 | 75.00% |

**Table 3: Distribution of study participants based on surgical outcomes (n = 28)**

| Outcomes | Category | Frequency (n) | Percentage (%) |
|---|---|---|---|
| **Surgery Performed** | Segmental Resection + Anastomosis | 25 | 89.30% |
| | Polypectomy | 3 | 10.70% |
| **Postoperative Complications** | Yes | 3 | 10.70% |
| | No | 25 | 89.30% |
| **Recurrence** | Yes | 7 | 25.00% |
| | No | 21 | 75.00% |
| **Follow-up Duration (months)** | < 12 months | 11 | 39.30% |
| | ≥ 12 months | 17 | 60.70% |

**Table 4: Comparison of variables between syndromic and non-syndromic cases (n = 28)**

| Variable | Syndromic n (%) | Non-Syndromic n (%) | Test Statistic (Fisher's) | p-value |
|---|---|---|---|---|
| **Sex (Female)** | 7 (63.6) | 11 (64.7) | 0.000 | 1.0000 |
| **Vomiting** | 11 (100.0) | 10 (58.8) | 5.200 | **0.0300*** |
| **Family History** | 11 (100.0) | 0 (0.0) | 28.000 | **<0.0001*** |
| **Multiple Polyps** | 11 (100.0) | 0 (0.0) | 28.000 | **<0.0001*** |
| **Recurrence** | 7 (63.6) | 0 (0.0) | 9.100 | **0.0050*** |

**Fisher's Exact test*

## DISCUSSION

The present study distinguishes the clinical and pathological features of syndromic versus non-syndromic jejunojejunal

 

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intussusception in adolescents, highlighting key differences that impact diagnosis, surgical management, and long-term follow-up. Syndromic cases were significantly associated with positive family history and the presence of multiple polyps, underscoring a likely genetic etiology. Notably, all syndromic cases demonstrated either juvenile or hamartomatous polyps, consistent with Peutz–Jeghers or juvenile polyposis syndromes, while non-syndromic cases typically involved solitary lesions without systemic features. This finding is consistent with Attard et al., who emphasized the importance of early endoscopic surveillance in children with polyposis syndromes due to the high risk of obstruction and intussusception. [1] Similarly, Jelsig et al. documented early-onset intussusception in patients with Peutz–Jeghers syndrome, stressing the need for timely polyp detection and resection to prevent recurrence. [2] Our data reinforce the role of structured surveillance programs in syndromic polyposis. Sandru et al. described endocrine and cutaneous manifestations in syndromic patients, corroborating the multisystem involvement observed in our cohort and underscoring the need for multidisciplinary care.[3] Valle and Monahan provided a framework for classifying hereditary GI polyposis syndromes, highlighting the diagnostic value of family history and genetic testing.[4] This supports the incorporation of targeted genetic counseling and testing in patients with suggestive clinical features, enabling both early diagnosis and family screening.

Srivastava emphasized that even subtle histologic findings should prompt evaluation for inherited syndromes, especially in the presence of multiple polyps.[5] Castori et al. expanded this perspective by linking connective tissue disorders and GI presentations, broadening the phenotypic scope relevant to syndromic diagnosis.[6] In line with these, both Attard et al.[1] and Valle[4] recommended long-term surveillance, which is consistent with our finding of seven recurrences, all within the syndromic group. Regular endoscopic follow-up is therefore critical for managing long-term outcomes. Dieterle et al. noted that while non-syndromic cases can present with intussusception, they usually lack systemic features and tend to follow a more benign course, as reflected in our non-syndromic subgroup. [7] Carrim et al. similarly described isolated juvenile polyps as benign causes of intussusception in children without genetic syndromes, aligning with the conservative management observed in non-syndromic patients. [8]

On the other hand, Tam et al. highlighted diagnostic complexities in phenotypically variable syndromes, a challenge also encountered in differentiating syndromic versus sporadic GI disease. [9] Fölster-Holst et al. demonstrated the diagnostic importance of mucosal changes in genodermatoses, reinforcing the value of extraintestinal signs in identifying syndromic cases. [10] However, discrepancies exist. Some pediatric studies have shown recurrence and obstruction even in non-syndromic cases, suggesting that pathology alone may sometimes mimic syndromic disease.[11] In other reports, genetic testing was negative despite clinical features suggestive of a syndrome, raising questions about variable gene penetrance and unidentified mutations.[12] Moreover, emerging data indicate that non-syndromic juvenile polyps may occasionally present as multifocal, particularly in inflammatory conditions, complicating classification.[13]

These inconsistencies underscore the limitations of using clinical presentation alone for classification and highlight the need for standardized diagnostic algorithms integrating histopathology, imaging, clinical criteria, and genetic testing. Despite the relatively small sample size, our study offers valuable insights by directly comparing syndromic and non-syndromic subgroups within a uniform surgical cohort. Such classification is not merely academic but has direct implications for therapeutic planning, long-term surveillance, familial risk assessment, and overall quality of life for affected adolescents.

## CONCLUSION

The present study demonstrates that syndromic jejunojejunal intussusception in adolescents is strongly associated with a positive family history and the presence of multiple polyps, whereas non-syndromic cases are more often characterized by solitary juvenile polyps without systemic involvement. These findings, in line with existing literature, underscore the importance of detailed history-taking, careful intraoperative assessment, and selective genetic evaluation in patients with suggestive clinical features. Although surgical outcomes were favorable in both groups, the significantly higher recurrence rate among syndromic patients highlights the necessity of structured long-term follow-up. Accordingly, all adolescent cases should be systematically evaluated for syndromic indicators, with histopathological examination of every resected lesion, and genetic testing offered to those fulfilling clinical or pathological criteria. Syndromic patients should be enrolled in surveillance programs to facilitate early detection and management of recurrent lesions. A multidisciplinary approach involving pediatric surgeons, gastroenterologists, geneticists, and pathologists is vital, and the creation of dedicated institutional or national registries would strengthen understanding of epidemiology, recurrence patterns, and long-term outcomes.

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