Spectrum of general surgical problems in the developmentally disabled adults

Kamran Khalid, *FRCSI, FRCSEd,* *Saleh M. Al-Salamah,* *MBBS, FRCS.*

ABSTRACT

Objectives: This study highlights the spectrum of general surgical problems necessitating admission on intellectually disabled adult patients. Problems encountered in the management and the ways to overcome various difficulties are highlighted.

Methods: Prospective collection of data on 63 consecutive developmentally disabled adult patients admitted to the Department of General Surgery, Riyadh Medical Complex (RMC), Riyadh, Kingdom of Saudi Arabia for various indications from January 2000 through December 2004. Demographic details, clinical presentation, diagnostic modalities, associated physical and neurological disabilities, coexisting medical condition, treatment options, morbidity and mortality were analyzed. Various difficulties encountered during the management and mean to overcome these problems are addressed.

Results: Sixty-three patients accounted for 71 admissions. Mean age was 26.7 years with a male preponderance (4.25:1). Fifty-four patients were admitted for various emergency conditions. History of pica could be obtained in 33% of the cases. Twenty-seven patients were admitted for acute abdomen. Volvulus of the colon (22.2%) and pseudo-obstruction (18.5%) were the most common causes of acute abdomen. Twenty-one patients were admitted with upper gastrointestinal bleeding. Reflux esophagitis was the most common cause of bleeding (62%). Overall morbidity was 41% for emergency admissions and 22% for elective surgery. Hospital mortality was 21.4% for emergency surgery. There was no death in elective cases.

Conclusion: Developmentally disabled patients comprise a special class of patients with peculiar management problems. The treating clinician should be aware of various unexpected conditions not found as frequently in the normal patient population. Apparent lack of pain does not exclude an acute emergency. Possible surgical condition should be suspected if there is vomiting, abdominal distension, fever, increased irritability of recent onset. Male gender and history of pica are added risk factors.

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Developmentally disabled patients pose peculiar management problems when presented with an acute surgical condition necessitating admission and possible surgery. Most are bedridden and have varying degrees of associated musculoskeletal and neurological disabilities. The clinician's decision to operate or not, depends upon a good history and adequate physical examination, relevant laboratory data and imaging investigations and awareness on the possible problem. All these factors are altered to a variable extent in the mentally retarded patients. Lack of communication, abnormal behavior, associated

From the Department of General Surgery (Khalid, Al-Salamah), College of Medicine, King Saud University Unit, Riyadh Medical Complex, Riyadh, *Kingdom of Saudi Arabia*.

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Address correspondence and reprint request to: Dr. Saleh M. Al-Salamah, Department of General Surgery, College of Medicine, King Saud University Unit, Riyadh Medical Complex, Riyadh, PO Box 31168, Riyadh 11497, *Kingdom of Saudi Arabia*. Tel. +966 (1) 4671585. Fax. +966 (1) 4679493. E-mail: smsalamah@hotmail.com

congenital anomalies and skeletal deformities, varying degrees of motor and sensory impairments, coexisting medical conditions, altered reaction to pain and medication and difficulties in the postoperative management make this special group of patients increasingly susceptible to late diagnosis and higher morbidity and mortality rates. The prevalence of mentally retarded children aged up to 18 years was observed to be 8.9 per 1000 children in the Kingdom of Saudi Arabia (KSA) in a population-based national survey conducted during 1996-1999.[1]

Riyadh Medical Complex (RMC) is a tertiary referral center and teaching hospital with 1400 beds. The Department of General Surgery, RMC includes an additional well equipped 6 bed Gastro Intestinal Bleeding Unit (GIBU) for the emergency management of all patients with acute gastrointestinal bleeding in close collaboration with the Department of Gastroenterology. This prospective study carried over 5 years period presents the spectrum of surgical problems in the adult intellectually disabled patients and details the outcome of their management. Different measures to overcome various difficulties in the surgical management of this high-risk patient population are discussed further.

Methods. This study is a prospective collection of data on 63 developmentally handicapped adult patients accounted for 71 admissions to the Department of General Surgery, RMC, Riyadh, KSA from January 2000 through December 2004. Patients were either referred from various rehabilitation centers for the "Mentally Retarded and Developmentally Disabled" in the Riyadh Region or from various departments of RMC. Mental retardation was defined by employing Diagnostic and Statistical Manual of Mental Disorders of the American Psychiatric Association, whereas the degree of retardation (mild; IQ between 50-55 and 70-75, moderate; IQ between 35-40 and 50-55, severe; IQ between 20-25 and 35-40, and profound; IQ of less than 20-25) was classified according to the Classification Manual of Mental Retardation of the American Association on Mental Deficiency.[2] The parameters studied were demographic data, duration of symptoms, clinical features, associated musculoskeletal and neurological disabilities, coexisting medical conditions, diagnostic modalities, operative findings, treatment and outcome in terms of morbidity and mortality. Various perioperative difficulties encountered in the management of these patients were noted. A protocol was developed to overcome these difficulties. X-rays of the abdomen were obtained in all cases presenting with feature of acute abdomen. Ultrasound and CT scan were other imaging modalities employed in elective and emergency cases, where a clinical diagnosis could not be made. Patients with gastrointestinal (GI) bleeding were admitted to GIBU and underwent emergency upper GI endoscopy (within 12 hours of admission). All remaining patients were admitted to the acute surgical ward with attention to frequent clinical assessment and consideration was made for subsequent high dependency (HDU) or intensive care units (ICU). The concurrent medical conditions were controlled, and anesthesiologist's consultation was sorted earlier. All patients requiring major surgical procedures were managed postoperatively in HDU or ICU until stabilized. Postoperative complications were recorded. The hospital mortality was defined as death within the same admission or 30 days of hospital stay. The surviving patients were discharged back to the referring rehabilitation centers or respective wards for continued care, with an advice for regular outpatients follow up every month for the first 6 months, every 3 months for a period of 2 years and as required thereafter. Patients with stomas were readmitted electively after 8-12 weeks for subsequent closures. Patients with GI bleeding had regular follow up in GI clinic.

Results. Twenty-seven (43%) patients presented with clinical features of the acute abdomen, 21 (33%) were admitted with acute upper gastrointestinal bleeding and an additional 15 (24%) patients were admitted with non-abdominal emergencies (6 cases) or for elective procedures (9 cases). There were 51 (81%) males and 12 (19%) females with male to female ratio of 4.25:1. The mean age was 26.7 years (range, 16-39 years). Degree of retardation on 19 (30%) patients was categorized as "profound", 17 (27%) as "severe", 14 (22%) as "moderate" and 13 (21%) as "mild". Out of the 54 cases admitted as emergency, the mean duration of symptoms was 1.8 (±1) days (range, 1-5 days) and 71% patients presented with a short history of less than 2 days. Thirty-four (54%) patients had varying degrees of associated physical abnormalities and another 19 (30%) had various unclassified neurological disabilities. Concomitant medical conditions were observed in 17 (27%) cases. Epilepsy was the most common associated disorder recorded in 9 patients, followed by diabetes (5 cases), congenital heart diseases (5 cases), chronic pulmonary diseases (4 cases) and severe anemia (3 cases). Twenty-three (36.5%) patients had severe and 18 (28.6%) had moderate malnutrition at initial presentation. Seven (11%) patients were receiving nasogastric and 4 (6%) were on gastrostomy feeding. History of pica could be obtained in 21 (33%) cases. The laboratory

investigations revealed mean hemoglobin of 9.3 gm/dL (range 7.8-11.4 gm/dL) and a mean albumen of 2.9 gm (range 2.3-3.5 gm) reflecting an overall low nutrition status. Eleven patients (17%) had grossly impaired renal functions suggestive of chronic renal impairment.

Out of 27 cases admitted for various acute abdominal conditions, 5 were managed successfully with various conservative measures, whereas 22 underwent laparotomy. There were 20 men and 7 women (2.8:1). Anorexia (refusal to take food) and vomiting of varying frequency were observed in all cases (100%) with acute abdomen, followed by abdominal distension (76%), pain (58%), and absolute constipation (51%). Abdominal tenderness (19 cases, 70%) was the most common clinical finding followed by guarding and rigidity (16 cases, 59%), abdominal distension (15 cases, 55.5%) and fever (11 cases, 41%). Plain x-rays of the abdomen, obtained in all acute cases were supportive of diagnosis in 12 (44%). **Figure 1** shows massively distended large bowel in a case of pseudo-obstruction of the colon. A CT scan was requested in 16 patients and was conclusive in 13 cases (81% accuracy). The scan remained inadequate in the remaining 3 patients. Eleven patients underwent colonoscopy, which was diagnostic in 6 (pseudo-obstruction 3, sigmoid volvulus 2, cecal volvulus 1) and suggestive in 5 cases (pseudo-obstruction 2, sigmoid volvulus 1, cecal volvulus 2). The examination, however, had to be abandoned or remained inadequate in 3 patients due to difficulty in maintaining adequate position, irritability and inadequate preparation. Two patients with pseudo-obstruction of colon and one each of sigmoid and cecal volvulus were managed successfully with colonoscopy. The remaining 7 patients were subsequently operated due to worsening abdominal signs. Acute intestinal obstruction remained the most common indication of surgery noticed in 4 (19%) cases, followed by acute appendicitis and pseudo-obstruction of large bowel observed in each 3 patients (14.3%). One female patient presented with hematemesis with features of acute abdomen. Upper GI endoscopy revealed a large leather foreign body penetrating the greater curvature of stomach. The patient underwent operative removal of the foreign body and repair of gastric perforation.

Twenty-one patients accounted for 27 admissions for upper GI bleeding during the study period. Nineteen were males and 2 were females, showing marked male preponderance (9.5:1). Fifteen (71%) were categorized as "profoundly" or "severely" retarded and 13 (62%) had associated physical disabilities of varying severity. The bleeding was usually mild and no patient needed surgical intervention. Whereas

Figure 1 - X-ray of plain abdomen showing massively distended large bowel (pseudo-obstruction of the colon).

15 of 21 patients did not require any transfusion, the average transfusion requirement for those who received blood was 1.5 Units. Reflux esophagitis was the most common cause observed in 13 (62%) cases admitted with hematemesis or malena. Associated hiatus hernia was reported in 9 of these patients and 2 (15%) patients had developed Barrett's change. Four (19%) patients had erosive gastritis with or without erosive duodenitis. Bleeding duodenal ulcer was reported in 2 other cases. Severe erosive esophageal candidiasis and Dieulafoy's lesion were the source of bleeding in remaining 2 cases. All were managed successfully with endoscopic treatment followed by proton pump inhibitor (omeprazole®) therapy.

Of the 9 patients admitted for elective surgery, 3 underwent mesh repair of groin hernia, 2 had laparoscopic cholecystectomy and anatomical repair of paraumbilical hernia. One female patient was operated for multinodular goiter with retrosternal extension causing pressure symptoms. **Table 1** details various conditions necessitating admission in 63 cases.

Overall postoperative morbidity was 41% for emergency and 22% for elective cases. Chest infection (30.5%) was the most common complication followed by wound infection (22%). **Table 2** summarizes various postoperative complications. One patient who underwent right hemicolectomy for cecal volvulus was operated 3 times for wound dehiscence and anastomotic leak. He ultimately died of uncontrolled sepsis and multiorgan failure. The overall hospital mortality for 28 patients operated in emergency (22 abdominal, 6 non-abdominal) was 21.4% (6 cases) and nil for elective surgery. Four out of 5 patients

Table 1 - Diagnosis, treatment and outcome in terms of mortality in 63 patients.

Diagnosis	n (%)*	Operation (n)	Outcome
Acute abdomen	**27**		
Large bowel pseudo-obstruction	5 (18.5)	Cecostomy (3), Colonoscopic decompression (2)	1 Died
Acute Small bowel obstruction	4 (14.8)	Congenital band (2), adhesive (1), tuberculous (1)	
Acute appendicitis	4 (14.8)	Appendicectomy (4)	
Sigmoid volvulus	3 (11.1)	Sigmoid colectomy (2), Colonoscopic untwisting (1)	1 Died
Cecal volvulus	3 (11.1)	Right colectomy (2),Colonoscopic untwisting (1)	1 Died
Small bowel perforation	2 (7.4)	Primary closure (1), Segmental resection (1)	
Meckel's diverticulitis	1 (3.7)	Wedge resection	
Foreign body stomach	1 (3.7)	Gastrotomy, removal and repair of perforation	
Gangrenous cholecystitis	1 (3.7)	Cholecystectomy	1 Died
Incarcerated paraumbilical hernia	1 (3.7)	Repair	
Mesenteric vascular occlusion	1 (3.7)	Extensive small bowel resection	1 Died
Traumatic rectal perforation	1 (3.7)	Repair with proximal diverting colostomy	
Non-abdominal emergency	**6**		
Abscess / carbuncle	3 (50)	Drainage/Debridement	
Strangulated hemorrhoids	1 (16.6)	Hemorrhoidectomy	
Diabetic gangrene foot	1 (16.6)	Below-knee amputation	
Necrotizing fascitis perineum	1 (16.6)	Repeated debridement and excision	1 Died
Upper GI bleeding	**21**		
Reflux esophagitis	13 (61.9)	Endoscopic treatment + Proton pump inhibitors	
Erosive gastritis and duodenitis	4 (19)	Proton pump inhibitors	
Bleeding duodenal ulcer	2 (9.5)	Endoscopic heat probe coagulation	
Dieulafoy's lesion	1 (4.8)	Endoscopic heat probe coagulation	
Severe esophageal candidiasis	1 (4.8)	Local and systemic antifungal	
Elective	**9**		
Inguinal hernia	3 (33.3)	Repair	
Cholecystecomy	2 (22.2)	Laparoscopic cholecystectomy	
Paraumbilical hernia	2 (22.2)	Repair	
Retrosternal goitre	1 (16.6)	Subtotal thyroidectomy	
Pilonidal sinus	1 (16.6)	Excision	
Total	**63**		**6 died**

*Percentage are calculated for respective number of patients in each group.

Table 2 - Complications observed in operated patients (n=36).

Complication**	Emergency n=27*		Elective n=9	
	n	(%)	n	(%)
Chest infection	9	(33.3)	2	(22.2)
Wound infection	7	(25.9)	1	(11.1)
Fatal septicemia	5	(18.5)	-	--
Abdominal collections***	3	(14.3)	-	--
Urinary tract infection	4	(14.8)	1	(11.1)
Seroma	3	(11.1)	1	(11.1)
Delayed wound healing	4	(14.8)	1	(11.1)
Wound dehiscence***	1	(4.8)	-	--
Anastomotic leaks***	1	(4.8)	-	--
Massive pulmonary embolism	1	(3.7)	-	--

*one patient died in the recovery; **several patients had multiple complications; ***percentages calculated for patients undergoing abdominal surgery (n=21)

who died, presented late (more than 72 hours) with features of advance sepsis following acute abdomen, and 3 of them had underlying colonic pathologies (volvulus, 2 cases, pseudo-obstruction 1 case). One additional diabetic patient with severe necrotizing fascitis of perineum died of uncontrolled sepsis.

During the mean follow up of 3.2 years, no procedure related late complication necessitating second surgery was observed. One patient developed an incisional hernia through scar of colostomy closure. One patient who initially underwent segmental small bowel resection for tuberculous enteritis, presented twice with subacute intestinal obstruction, but was managed successfully with conservative treatment. Patients with upper GI bleeding had a regular follow up in the GI outpatients' clinic. Two patients with Barrett's change are under continued surveillance with repeated endoscopies and biopsy every 6 months.

Discussion. Developmentally disabled patients pose clinical and management challenges, especially when presenting with various conditions requiring surgical intervention. Intellectual disability (mentally handicapped, mentally retarded, developmentally disabled) is defined as: significantly sub average intellectual functioning; existing concurrently with related limitations in 2 or more of the adaptive skills (communication, self-care, home living, social skills, use of community resources, functional academic skills, health and safety, leisure and work); manifested before the age of 18 years.[2,3] Previous study observed the prevalence of mentally retarded children aged up to 18 years to be 8.9 per 1000 children in a population-based national survey and was well correlated with 6.2-12 per 1000 population reported from various developed countries.[1] The etiological factors leading to mental retardation vary across different populations, but a strong association with low socioeconomic status has been observed.[1-4]

This study, details the experience of a tertiary referral teaching hospital from Central Riyadh in the management of this special patient population requiring surgery for various emergency and elective conditions. As all patients with GI bleeding are admitted to GIBU of the Department of General Surgery in RMC and managed initially by the surgical team with early endoscopy by the gastroenterologist, following acute management, these cases are also included in the overall spectrum of general surgical admissions in this study.

The higher male to female ratio (4.25:1) and younger age (mean 26.7 years) are similar to other reports.[5-7] Most of the patients had a short history of less than 48 hours. Anorexia, vomiting and abdominal distension were the most common presenting features for acute abdominal emergencies. These observations are in accordance with various other studies.[5,7] We obtained history of pica in 33% of our cases. Five of 8 patients with history of pica in this study presented late with more than 3 days history of acute abdominal complaints and 2 (25%) of them died. This confirms the ominous nature of pica in this group of patients. In his study of 25 mentally retarded patients, Voitk[5] in 1987 quoted deaths from pica at 33% and reported history of pica in 40% of patients presenting with acute abdomen as against 16% in the reference population. The author observed pica as a cause of repeated admissions, delayed presentation and higher (25%) mortality. Volvulus of sigmoid colon or cecum was the most common cause of acute abdomen observed in 22.2% of cases followed by pseudo-obstruction of large bowel (18.5%). This signifies this group of patients as increasingly susceptible to such conditions. Of special mention is cecal volvulus (11.1%), the incidence of which seems to be higher than in normal population. The clinician should be aware of such unexpected conditions whenever faced with mentally handicapped patients presenting with acute abdomen and colonic distension. Every effort should be made for early colonoscopic diagnosis and treatment to avoid higher morbidity and mortality related to late diagnosis. Accordingly, 2 of 6 patients with volvulus of colon and 2 of 5 patients with colonic pseudo-obstruction underwent successful colonoscopic management in the present study, whereas 3 of the 7 operated patients who presented late died subsequently (42.8% mortality for colonic emergencies). Failure of repeated colonoscopic decompressions or incomplete or inadequate procedure on account of fecal loading or impaction, improper positioning or lack of cooperation should lead the clinician to keep a low threshold for an early exploration in the mentally retarded patients. Various other unexpected conditions, not as frequently observed in the normal population, include foreign body ingestion resulting in bowel obstruction or perforation and traumatic rectal perforation makes recognition of the acute abdomen, and decision for subsequent surgery, in the mentally retarded patients difficult for a surgeon who is not accustomed to these diagnoses in the general population. The altered reaction to pain, concomitant use of various medications, concurrent medical conditions, difficulty in communication, malnutrition and associated neurological and musculoskeletal disabilities and deformities also contribute to delayed recognition. Abnormal gas patterns observed in abdominal x-rays are often considered normal in such patients and make interpretation difficult.[6] Moreover, the musculoskeletal deformities may make various imaging studies, like ultrasound, CT or contrast studies, difficult to obtain and interpret, thus adding to the diagnostic difficulties. The high morbidity and mortality figures are directly associated with the delayed diagnosis and various other management difficulties encountered in these unique high risk patients. Accordingly, we recorded high overall morbidity (41%) and hospital mortality (21.4%) for emergency operation in this study. These figures are in accordance with various other reported series.[5-7] Higher rates of chest and wound complications are reflection of overall management problems like malnutrition, inadequate nursing care of the patient due to associated deformities and poor hygiene status. Various measures to overcome all these problems lead to the development of a departmental protocol to minimize these management difficulties and to reduce the associated higher morbidity and mortality rates.

Figure 2 - Postoperative management of patient in intensive care unit (note the possible need for ventilatory support due to chest deformities and nursing difficulties associated with contractures and skeletal deformities)

These include: a high index of suspicion for such conditions in the mentally retarded presenting specially with anorexia, vomiting, abdominal distension, prolonged constipation, fever or increasing irritability of recent origin, early admission to an acute surgical ward rather than continued observation in emergency department, frequent physical examinations and judicious use of tranquilizing medications, correction of any existing fluid and electrolytes abnormalities and adequate control of co-morbid medical conditions. Restraints may be used if necessary. Postoperatively, the patients should be managed in the acute ward, high dependency or intensive care units with close monitoring until stabilized and recovered (**Figure 2**). Tranquilizing drugs and restraints may be used to overcome agitation, and removal of various vital tubes, catheters and intravenous lines. The enteral feeding should be started early and every effort should be made towards rapid ambulation.

High male preponderance (9.5:1) and increased incidence of reflux esophagitis (62%) in patients admitted with GI bleeding is consistent with reports by other authors.[8-10] The high prevalence (15%) of Barrett's esophagus noticed in this study is in accordance with various other published studies in the mentally retarded.[8-11] We noticed that male gender, hiatus hernia, increasing severity of retardation and associated verbal and physical disabilities are significant risk factors for GI bleeding in the developmentally disabled population. These observations are in accordance with various other authors.

In conclusion, developmentally disabled patients comprise a special class of patients with peculiar management problems. The treating clinician should be aware of various unexpected conditions not found as frequently in the normal patient population. The apparent lack of pain does not exclude an acute emergency. Possible surgical condition should be suspected if there is vomiting, abdominal distension, fever, increased irritability of recent onset. Younger age, male gender and history of pica are added risk factors. We recommend frequent clinical examination and a low threshold for an early operation to decrease the associated high morbidity and mortality.

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