Factors that predict mortality of jejunoileal atresia in neonates in Baghdad2010-2012

Amer Ejrish Alkafaji
College of Medicin, University of Babylon
amierejrish@yahoo.com

Abstract

Background : Jejunoileal atresia and stenosis are the most common congenital anomalies of the small intestine and are a major cause of intestinal obstruction in neonates. Late intrauterine mesenteric vascular occlusions is the most accepted hypothesis as an etiology of jejunoilealatresia .

Objective : To describe the mortality rate of 64 patients with intestinal atresia between October 2010 and September 2012 and submitting initial scoring system of 50 points based on the estimated risk scores and P.value of each risk factors.

Methods :A prospective study of mortality rate in jejunoileal atresia according to the patients' parameters (prenatal diagnosis, prematurity , birth weight , age and severity of presentation , associated anomalies ,anatomic location and type of atresias,surgical techniques, early postoperative complications and re-interventions) .

Results : There were 64patients with jejunoileal atresia (35 male). Prenatal diagnosis was established in 5 patients (8%). The mean birth weight was 2.6 kg and higher mortality rate with lower birth weight. The mean gestational age was 37 weeks and 20% of the patients were premature. Thirty seven patients were presented to medical services in the first three days of life with lowest mortality rate, patients who were presented later had rate of death of 27%-60%. Sepsis and/or pneumonia were found in (22%) of patients clinically diagnosed with mortality rate (43%). Seventeen patients (26.5%) had different associated anomalies with related mortality of (53%). The highest mortality was related to high jejunal atresia (50%). Apple-peel & multiple atresias variants of jejunoileal atresia had the highest rates of mortality, 75% &43% respectively. The mortality rate of the 40 patients with end to obligue anastomosis with plication of proximal atretic dilated segment was 12.5%, while it was 58 % to that without plication of proximal atretic dilated segment. Nineteen patients (29.5%) were presented with early postoperative complications. Anastomotic complications (leak & non–functioning anastomosis) & short bowel syndrome were the major contributor to the related mortality rate (58%) .A surgical re-exploration was done in nine patients with high mortality rate (55.5%).The overall mortality was 16/64 patients (25%).Estimated risk scores and P.values for each factors were obtained. Scoring system of 50 points was developed according to these above values .

Conclusion: The overall mortality rate in the current study was 25%. The higher mortality rate was associated with those who had major risk factors compared to the lowest mortality rate of those with minor or no risk factors. According to estimated risk scores and P.values, cumulative scoring system was submitted .

Late presentation,late diagnosis, absence of well trained neonatal intensive care personnel and unavailability parenteral nutritional support were the major problems that contributed to this high death rate .

Keywords: jejeno-ileal , atresia, anastometic leak.

الخلاصة

خلفية عن الموضوع :ضمور وتضيق الامعاء الدقيقة الولادي هو من اكثر التشوهات الخلقية شيوعا والسبب الاساسي لانسدادات الامعاء الولادية. انسداد الاوعية الدموية المساريقية في الفترة الاخيرة من الحمل هو النظرية الاكثر قبولا كمسبب للمرض .

الاهداف :وصف نسبة الوفاة لحالات ضمور الامعاء الدقيقة الولادي عند 64 مريض , للفترة من 2010 الى 2012 مع تقديم نظام جمع اولي للنقاط يتكون من 50 نقطه معتمدا على نقاط الخطورة المقدرة لكل عامل خطورة .

الطرق : الدراسة تتضمن حساب نسب الوفيات لحالات ضمور الامعاء الدقيقه الولادي وفقا لعوامل الخطوره الرافقه للمرض وتحديدا : التشخيص قبل الولادة, الخداج, الوزن عند الولادة, العمر عند المراجعة الاولية من اجل العلاج, شدة الاعراض ,التشوهات الاخرى, الموضع التشريحي و نوع الضمور في الامعاء, الطرق الجراحية المستخدمة في العلاج, عقابيل ما بعد العمليات الجراحية و اعادة التداخل الجراحي .

النتائج : الدراسة تتضمن اربعة وستون مريض يعاني من ضمور الامعاء الدقيقه الولادي, بضمنهم 35 مريض من الذكور. ثبت التشخيص ما قبل الولادة عند 5 من المرضى فقط (8%) .معدل الوزن لدى المرضى كان 2.6 كغم مع ملاحظة ان نسبة الوفاة الاعلى كانت عند المرضى ذوي الاوزان القليلة. بالرغم من انمعدل فترة الحمل للمرضى كانت 37 اسبوعا ,فقد كانت نسبة الخداج هي20% . سبعه وثلاثون مريض قد حضروا الى المستشفى خلال الثلاثة ايام الاولى بعد الولادة حيث ان نسبة الوفاة كانت قليله بينهم مقارنة مع المرضى الذين راجعوا بعد هذه الفترة, حيث تراوحت

نسبة الوفاة لديهم 26 –60 % .حالات تعفن الدم المصاحبة للمرض هي 22% ,وكانت نسبة الوفاة بينهم 43% تقريبا .سبعة عشر مريض كان يعاني من تشوهات مختلفة اخرى ونسبة الوفاة المتعلقة بهم هي 53% .نسبة الوفيات الاعلى كانت ضمن المرضى اللذين يعانون من ضمور الامعاء في منطقة المعي الصائم وكانت تقريبا 50% .الانواع المعقدة و المتعددة من حالات ضمور الامعاء الولادي كانت الاكثر بين نسب الوفيات وهي 75% و43% على التوالي .

نسبة الوفاة بين المرضى اللذين عولجوا بعمليات جراحيه متضمنة ربط الامعاء مع طي الامعاء المتسعة كانت 12,5%, بينما كانت 58% للمرضى اللذين عولجوا بنفس الطريقة ولكن بدون طي الامعاء .

تسعة عشر من المرضى عانوا من عقابيل ما بعد العملية الجراحية. اعادة العملية الجراحية كانت ضرورية عند تسعه من المرضى مع ارتفاع نسبة الوفاة بينهم (5,.55%).

النسبة العامة للوفاة في هذه الدراسة المتكونة من 64 مريضا كانت 25% (16 مريض) .

في هذه الدراسة, تم استخراج نقاط الخطورة المقدرة لكل عامل خطورة, ووفق ذلك وضع نظام يجمع نقاط الخطورة مكون من 50 نقطه .

الاستنتاجات : النسبة العامة للوفاة في هذه الدراسة المتكونة من 64 مريض كانت 25% (16 مريض) .نسب الوفيات الاعلى كانت ضمن المرضى اللذي لديهم عوامل الخطورة العالية او الاساسية مقارنة مع هؤلاء المرضى مع عوامل الخطوة الثانوية او الذين ليس لديهم اي عامل خطورة .تطوير نظام لجمع نقاط الخطورة لا يزال يحتاج الى سريان للمفعول وتدقيق لصحته من خلال الدراسات المستقبلية المتعددة في اكثر من مركز تعليمي متضمنا اعدادا اكثر للمرضى .

التكهن بحصيلة هذا المرض تغير تغيرا جذريا خلال العقود الاخيرة في الدول المتقدمة, ولكنفي الدول النامية ومن ضمنها دولتنا ,لا زالت نسبة الوفيات مرتفعه مقارنة مع ما ورد في الدراسات والمناهج المطبوعة.

قد تكون اهم الاسباب التي تعزى لارتفاع نسبة الوفيات هي : تأخر المريض باللجوء الى المستشفى, تأخر تشخيص المرض ,غياب الاشخاص المدربين للعناية المركزة بحديثي الولادة والخدج, اضافة الى عدم توفر محاليل التغذية الوريدية الضرورية بشكل اساسي في مستقبل هذا المرض .

الكلمات المفتاحية: انسداد الصائم واللفائفي، التسريب من الربط

Introduction

Jejuno-ileal atresia and stenosis are the most common congenital anomalies of the small intestine (2. 9 cases per 10, 000 live births) and are a major cause of intestinal obstruction in neonates.(James *et al.*, 2006; Arnold & Scott, 2012) .The most accepted etiological hypothesis is late intrauterine mesenteric vascular occlusions(Sadler,). The vascular and subsequent ischemic insult not only causes morphologic abnormalities but also adversely influences the structure and subsequent function of the remaining proximal and distal bowel (Lewis& Arnold, 2006; George *et al.*,2010).

Aims of the study

1. Proper determination of the mortality rate of neonates with jejunoileal atresia in our region is particularly important for comparative purposes with that of the developed countries.
2. Identifying the most important risk factors (major ,minor ,& no risk) that contributed to the mortality rate in jejunoileal atresia according to estimation of the risk scores .
3. Estimation of mortality rate of each risk groups distributed by the anatomical location of the jejunoileal atresia
4. Submitting initial scoring system of 50 points based on the estimated risk scores and P.value of each risk factors.

Patient and method

A prospective hospital based study of 64 neonates (35 male, 29 female) with Jejuno-ileal atresia were selected, all the patients were surgically managed at the Department of Pediatric Surgery / Welfare teaching hospital / Medical City / Baghdad / Iraq from october 2010 to September 2012. The patients were followed for 4 – 6 months and the outcomes were evaluated by mortality during this period . This study is supported by written and verbal consents of the patients' relatives & agreement of the hospital administration . Prenatal and postnatal risk factors were evaluated that may be predicted the mortality of these cases (table-1). Those cases were essentially clinically diagnosed that were supported by plain

abdominal x-ray (erect , supine or lateral decubitus) with or without contrast enema ,thereafter , they were adequately resuscitated under warm humidified environment (incubator) with intravenous fluids & antibiotics. All of those patients were surgically managed via several operative techniques by different surgeons. The surgical options involved in the management of these cases were either resection of the proximal & distal atretic bowel & then single layer end-to-oblique anastomosis with 4-0 absorbable suture material with or without anti-mesenteric plication , or enterostomy.

Data were transferred into a computerized data form , and analyzed by statistical package for social sciences (SPSS) software for windows version 20, IBM, US. The variables including the risk factors and gender were presented as frequencies and proportions (%). Age was presented as median and range. The outcome was categorized as died or survived and the mortality rate was calculated in relation to each risk factor.

The estimated risk for each factor was calculated depending on the odds ratio by cross tabulation of each risk factor against outcome. The risk factors were categorized according to the estimated risk into three categories; Major risk factors included the risk factors that have an estimated risk of more than 2 and or highly significant correlation with the outcome. Minor risk factors included the risk factors that own a significant estimated risk of more than 1 and less than 2, the third group included the risk factors that own an estimated risk of less than 1, and named as no risk factors.

Depending on the estimated risk, a scoring system developed of 50 points, which represented the cumulative estimated risk score for all risk factors after approximation of the estimated risk score for each factor to the nearest digit. Then patients were categorized into 4 groups according to their cumulative risk score; <10, 10-19, 20-30 and ≥ 30 score. These scores groups were cross-tabulated against outcome to find the correlation between the cumulative risk score values and the outcome

The significance of differences or correlation was estimated by using chi square test. P. value ≤ 0.05 was considered as significant. Results, finally, were presented in tables and figures.

Table-1 : Prenatal and postnatal risk factors of Jejuno-ileal atresias' patients .

Risk factors	
Prenatal factors (Prenatal ultrasonography)	**Polyhydraminos**
Postnatal factors	• **Prematurity**
	• **Gender**
	• **Birth weight**
	• **Age of presentation**
	• **The severity of presentation**
	• **Associated anomalies**
	• **Anatomic location of the atresia**
	• **Type of atresia**
	• **Surgical techniques**
	• **Reoperation**
	Postoperative need for NICU admission

Result

A total of 64 neonates with JIA were enrolled in this study, they were 35 males (54.7%) and 29 females (45.3%) with a male to female ratio of 1.2:1 . The median age at presentation was 3 days and the range of ages was (1 - 21) days . According to the estimated risk scores of each risk factor , the risk factors were categorized into three categories (Table-2).

As it is shown in (Table 4), patients with major risk factors were 36 patients with mortality rate of 38.9% , those with minor risks were 24 patients with only 2 patients (8.3%) died, while those with no risk were only 4 and all were survived .These findings indicated a significant direct correlation between the major risk factors and mortality, P=0.014.

The correlation between the mortality and risk categories regarding the anatomic location revealed that the higher mortality rate in patients who had jejunal site and major risk factors followed by those with minor risk factors while the higher survival rate was present in patients with no risk factor , this correlation was found also in other anatomic site, P<0.05 (table 5).

Depending on the risk score for each risk factor shown in (Table 3) and (table 6), the cumulative risk score of 50 points was calculated. The lower cumulative risk score in the studied group was 3, and the higher score was 33. The patients were categorized according to their cumulative risk score into four groups (Table 7); 36 patients had a risk score less than 10, those with risk score 10-19 were 21, six patients had a risk score of 20-29 and only one patient had a risk score more than 30, it had been significantly found that mortality rate was directly correlated to the score value; those with higher risk score had the higher mortality rates, out of the 36 patients with < 10 score only 3 patients (8.3%), among those with 10-19 risk score the mortality rate was 33.3%, to reach 83.3% in those with a score of 20-29, and the only one patient with a score of > 30 was dead (100% mortality), this indicated the significant positive correlation between the risk score and mortality, these findings were shown in(table 7) , (figure 2) and (figure 3).

Table-2: Categories of patients according to the estimated risk scores of each risk factor .

Major risk factors
Factors with estimated risk score (odds ratio) of more than two folds &/or with highly significant P –value
1. Birth weight of less than 1600 grams
2. Prematurity
3. Presentation to the medical services initially in the second week of life
4. Presentation with sepsis and/or pneumonia
5. Severe associated anomalies (biliary atresia ,total aganglionosis , lately presented gastroschisis , perinatal perforated viscus ,omphalocele major , severe urological & cardiac anomalies)
6. Type III b
7. Poor surgical techniques (delayed operation & resection and primary anastomosis with end to obligue anastomosis without plication of proximal atretic dilated segment)
8. Associated postoperative anastomotic complications and short bowel syndrome that need for reoperation
The estimated mortality range for this category was 38.9 %. The free survival rate was around 60 %.
Minor risk factors
Factors with estimated risk score (odds ratio) of one to two folds &/or significant P –value.
1. Birth weight of 1600 – 2400 grams
2. Presentation to the medical services in the first week of life

3. Moderately severe associated anomalies (Meckel 's diverticulum , small & early presented gastroschisis and omphalocele minor)
4. Type IV (not familial form) & Type III a
5. Temporary enterostomy and mucous fistula
6. Associated postoperative complications (mainly wound infection and stomal complications)
The estimated mortality range for this category was 8.3 %. The free survival rate was around 91%.
No risk factors
Factors with estimated risk score (odds ratio) of less than one fold with&/or not significant P –value
1. Birth weight of ≥ 2500 grams
2. Full term
3. Presentation to the medical services in the first three days of life
4. No associated sepsis and/or pneumonia on presentation
5. No associated anomalies
6. Type I & Type II
7. Optimal surgical techniques (early operation & primary resection and end to obligue anastomosis with placation of the proximally dilated segment)
8. No need for re-operation
9. No postoperative complications
The estimated mortality rate for this category was 0.0 %. The free survival rate approached 100%

Table 3. Correlation between risk factors and mortality among studied group with the estimated risk scores .

Patients' variables		Died (n=16)		Survived (n=48)		Estimated risk score (odds ratio)	P.value
		No.	%	No.	%		
Prenatal diagnosis	Yes	2	40%	3	60	1	0.78
	No	14	22%	45	78		
Gender	Male	9	25%	26	75	2	0.88
	Female	7	24%	22	76		
Weight (gm)	≤1500	3	18.8	3	6.2	3.5	0.011
	1600-2400	7	43.8	17	35.4	1.4	0.02
	≥ 2500	6	37.5	28	58.3	0.43	0.3
Gestational age	Preterm	7	43.8	6	12.5	5.4	0.007
	Full Term	9	56.2	42	87.5	0.4	0.012
Age of presentation	First 3 days of life	7	43.8	30	62.5	0.47	0.18
	3 - 7 days of life	6	37.5	16	33.3	1.2	0.76
	After 7 days of life	3	18.8	2	4.2	5.3	0.003
Presentation	With sepsis or pneumonia	6	37.5	8	16.7	3	0.04
	without sepsis or pneumonia	10	62.5	40	83.3	0.33	0.04

Site of Atresia	Jejunal	10	62.5	10	20.8	2.8	0.013
	Mid-small bowel	4	25.0	24	50.0	1	0.244
	Ileal	2	12.5	14	29.2	1	0.14
Associated anomalies		8	50.0	9	18.8	2.7	0.013
The Type of atresia	Type I	1	6.3	2	4.2	0.13	0.73
	Type II	2	12.5	11	22.9	0.87	0.14
	Type III a	7	43.8	30	62.5	1.1	0.19
	Type III b	3	18.8	1	2.1	2.5	0.02
	Type IV	3	18.8	4	8.3	1.4	0.017
Anastomosis	with Plication	5	31.2	39	81.2	0.2	<0.01
	without Plication	7	43.8	1	2.1	5.4	<0.01
Stoma		4	25.0	8	16.7	1.4	0.46
Early post operative complications		6	37.5	13	27.1	1.4	0.43
Reoperation		5	31.2	4	8.3	2.8	0.022

Table 4. Correlation between Risk group and mortality rate .

Risk group	Survive		Died		Total	
	No.	%	No.	%	No.	%
No risk	4	100.0	0	0.0	4	100.0
Minor risk	22	91.7	2	8.3	24	100.0
Major risk	22	61.1	14	38.9	36	100.0
Total	48	75.0	16	25.0	64	100.0

Table 5: Association between risk factors of the patients and the anatomic location of jejunoileal atresia regarding the mortality rate .

Risk category		Site					
		Jejunal		Mid-small bowel		Ileal	
		Survive	Died	Survive	Died	Survive	Died
No Risk	No.	2	0	2	0	0	0
	%	100.0	0.0	100.0	0.0	0.0	0.0
Minor Risk	No.	2	1	13	0	7	1
	%	66.7	33.3	100.0	0.0	87.5	12.5
Major Risk	No.	7	8	8	5	7	1
	%	46.7	53.3	61.5%	38.5	87.5	12.5
Total	No.	11	9	23	5	14	2
	%	55.0	45.0	82.1	17.9	87.5	12.5
P.value = 0.043							

Table 6. The estimated and cumulative risk score for all risk factors approximated to the nearest digit.

Patients' variables(risk factor)		Score
Weight (gm)	**<1500**	**4**
	1600-2400	**2**
	> 2500	**0**
Gestational age	**Preterm**	**6**
	Full Term	**0**
Age of presentation	**First 3 days of life**	**0**
	3 - 7 days of life	**2**
	After 7 days of life	**6**
Presentation	**With sepsis or pneumoni**	**4**
	without sepsis or pneumonia	**0**
Site of Atresia	**Jejunal**	**4**
	Mid-small bowel	**1**
	Ileal	**1**
Associated anomalies		**3**
The Type of atresia	**Type I**	**0**
	Type II	**0**
	Type III a	**0**
	Type III b	**3**
	Type IV	**2**
Anastomosis	**with Plication**	**0**
	without Plication	**5**
Stoma		**2**
Early post operative complications		**2**
Reoperation		**3**
Total risk score		**50**

Table 7. The risk score categories and associated outcome for each group .

Risk score category		Mort		Total
		Survive	Died	
< 10	**No.**	**33**	**3**	**36**
	%	**91.7**	**8.3**	**100.0**
10-19	**No.**	**14**	**7**	**21**
	%	**66.7**	**33.3**	**100.0**
20-29	**No.**	**1**	**5**	**6**
	%	**16.7**	**83.3**	**100.0**
≥30	**No.**	**0**	**1**	**1**
	%	**0.0**	**100.0**	**100.0**
Total	**No.**	**48**	**16**	**64**
	%	**75.0**	**25.0**	**100.0**

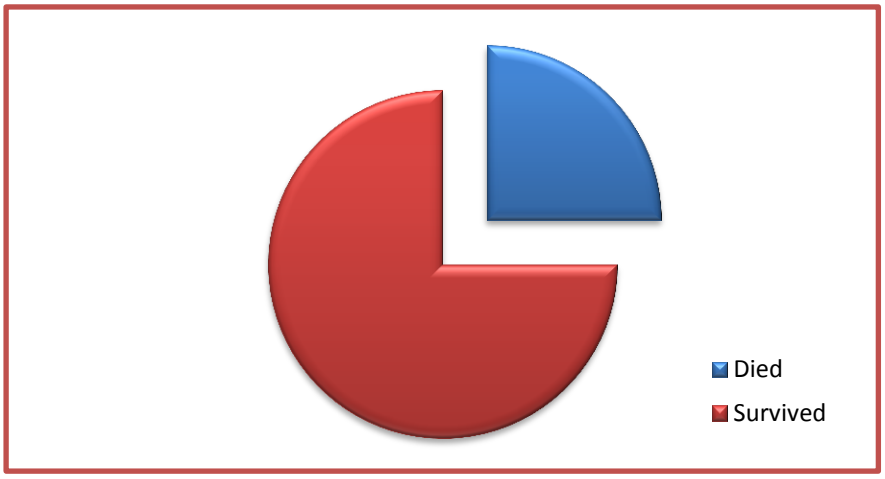

Figure 1. Distribution of outcome among studied group. (*N=64; Mortality rate was 25%)*

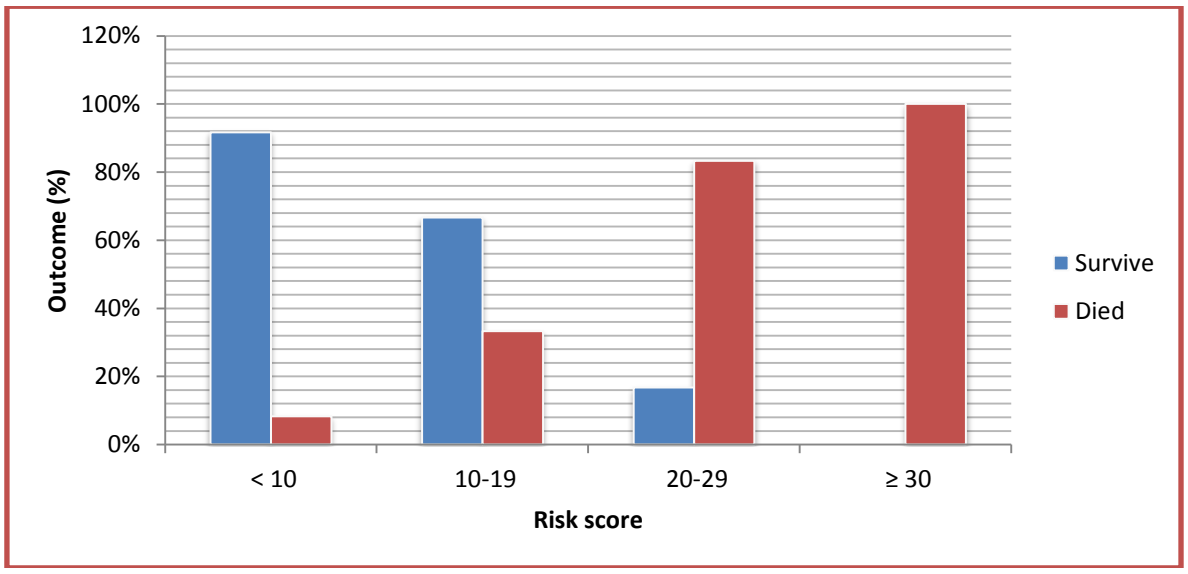

Figure 2 . Correlation between cumulative risk scores and outcome.

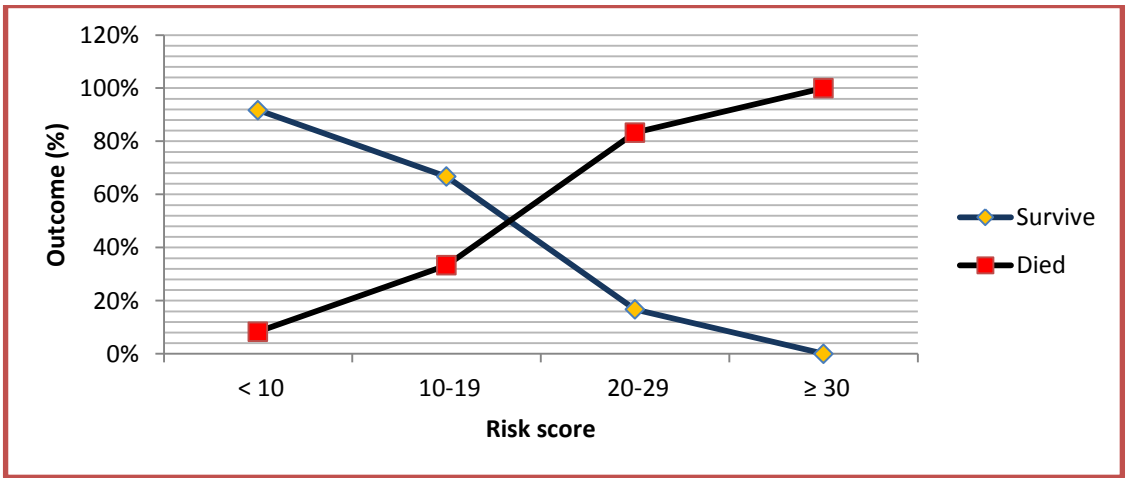

Figure 3. Trends of survival and mortality in relation to risk categories.

Discussion

Intestinal atresias are the most common cause of neonatal intestinal obstruction. . In our study , intestinal obstruction was suspected antenatally in 5 /64 (8%) compared to 15/47 (32%) in a study at 2002 in a regional pediatric surgical centre/ United Kingdom (Kumaran *et al.*,2002), while , no patient was diagnosed antenatally in a study of 28 patients by Shakya *et al*. in Nepal 2010 (Shakya *et al.*, 2010). Another study reported antenatal diagnosis in 41% (Stoll *et al.*, 1996).

The potential benefits of antenatal diagnosis include earlier recognition and parental counseling, prompt intervention and reducing metabolic complications (Nixon ,1971).

In our study, the limited number of those that antenatally diagnosed and the high associated mortality (40%) may be due to poor imaging studies (inexperience persons), poorly applied medical services, delayed referral, or poor family education regarding early presentation .

The gender distribution in this study was 35 male: 29 female ,with no significant difference in mortality rate of 25 % & 24% respectively . This agreed with other studies regarding the incidence and death rate ((Kumaran *et al.*, 2002; Shakya *et al.*, 2010; Thamar *et al.*, 2009) . The current study had median birth weight of patients of 2.6 kg. A study by Kumaran (Kumaran *et al.*, 2002) & other by Ekwunife (2011) argued that median birth weight of 2.82 kg & 2.3 kg respectively. Higher mortality rate was associated with lower birth weights especially in those who had proximal jejunal atresias that approached 50% in neonates with less than 1500 gram. This agreed with a study of Nixon and Tawes in 1971 ,but disagreed with an other study by Shakya (2010) ; (Shakya *et al.*, 2010) who stated no significant difference in death rate between jejunal and ileal atresias considering the sex ratio, age at presentation, birth weight , and frequency of complications .

The incidence of prematurity in our study was 20% with & mortality rate of 55% . Based on previous study (Thamar & Stollman 2009) in Netherlands) ,it is more than 50% of prematurity , but the overall mortality rate was 11%. This may be due to advanced management of premature patients in his region . The median gestational age of 37 weeks was compatible with other studies (Kumaran *et al.*,2002; Sathya ,2011).

The associated lower birth weights, low immunity, sepsis, metabolic abnormalities due to hypothermia and the added other congenital anomalies may be contributed to the higher mortality rates of prematurity.

Late Presentation in our study was associated with 60% mortality rate . Shakya *et al.* (2010) argued that overall delay in presentation has not been found to be a significant factor for mortality in those with incomplete obstruction, but in those with complete obstruction, it is still a significant factor for mortality .

In our study, 14 patients were presented with sepsis and/or pneumonia with mortality rate of 43%, this agreed with another study (Shakya *et al.*, 2010) which reported 5 patients with sepsis of 28 patients with jejunoileal atresia.

The high mortality rate in those patients with delayed presentation and related septic complications may be due to increased risk of intestinal perforations, nutritional & metabolic dearrangements.

Our recent study reported 20 patients with jejunal atresia , 28 patients with jejunoileal atresia and 16 patients with ileal atresia. Another study had higher incidence of jejunal atresias (Thamar & Ivode, 2009),while another one reported more ileal atresias (Shakya *et al.*, 2010). The above differences may be due to population variations.

The higher incidence of Type III(a) & the higher mortality rate with Type III(b) & Type IV were compatible between this study and another study (Thamar & Ivode, 2009).

The incidence of associated anomalies was 26.5%, which disagreed with the reported incidence of 52% in a study of 130 patients with intestinal atresias by Sarah Bouchard (Sathya, 2011). The incidence of cardiac anomalies in our study was only 2% when compared to 19 % in the above study (Sathya , 2011). This is may be poor screening for associated cardiac, urological and other anomalies in our medical services .

Lower mortality rate of 12.5% was reported with the optimal surgical techniques (early surgical intervention & primary resection and end to obligue anastomosis with plication of the proximally dilated segment), while that of poor surgical techniques (delayed surgical intervention & resection and primary anastomosis with end to obligue anastomosis without plication of proximal atretic dilated segment) may approach 58%. That agreed with another study regarding the effect of surgical techniques on outcome of patients with jejunoileal atresia (Shakya *et al.*, 2010; Dalla Vecchia *et al.,* 1998) .

In our study, the absence of neonatal intensive care also contributed to the mortality and morbidity. The anesthetic challenge, fraught with dearth of facilities and man power are closely related to this.

A lot of man hours were invested on very frequent manual observation of basic vital signs and the general state of the babies. In neonatal critical care, this is often inefficient with attendant fatigue and observer errors. On the other hand ,the high mortality rate (66.5%) of the 6 /64 patients who admitted to NICU , may be due to sepsis and/or pneumonia ,hospital acquiered infection and severe associated anomalies on presentation .that was completely similar to the result of a study in Nepal (Shakya *et al.*, 2010).

In our study, surgical re-intervention was done in 13.5% of the patients . Another study in Netherland in 2011 (Sathya, 2011) was obviously higher re-operation rate (25%), but both with high mortality rate.

Mortality has declined steadily throughout the 20th century. In the first half of the last century, mortality rate of patients with JIA was 90% (Evans, 1951). With improvements in neonatal anesthetic techniques, neonatal intensive care, operative techniques and TPN use, the survival figures climbed to 88% in the early 1990s (Kumaran *et al.*,2002).

Dalla Vecchia *et al.,* (1998) stated a long term survival of 84% in 128 patients with JIA in the period of 1972 to 1997 . In evaluating 30 cases of isolated JIA in 2002 , Kumaran *et al.,* 2002 observed that mortality rate had declined to nil . Kumaran, however, excluded patients with gastroschisis and their study refers to The period of 1991 to1998. Thamar H.Stollmana / Netherlands (Dalla Vecchia *et al.,* 1998) observed a late mortality rate of 11% in 114 patients in a period of 1971 - 2004. Another study proposed by Ahmed A. Khalaf at Welfare teaching hospital /Baghdad at 2009 , demonstrated mortality rate of 35% (Ahmed, 2010). The differences in mortality rates among variable studies ,in different regions of the world are shown in (table 8).

In our study, patients presented with no risk factors and atresia at high jejunum , mid-small bowel or distal ileal anatomic location had survival rate of 100% .

Patients presented with minor risk factors and atresia at high jejunum , mid-small bowel or distal ileal anatomic location had mortality rate of 33.3% , 0.0% and 12.5 % respectively .

Lastly , Patients presented with major risk factors and atresia at high jejunum ,mid-small bowel or distal ileal anatomic location had mortality rate of 53.3% , 38.5% and 12.5% respectively .

According to a risk stratification schema proposed by Nixon and Tawes in New York at 1971 (Nixon & Tawes, 1971) showed that patients with high jejunal atresia in group A (no risk factors) or B (minor risk factors) had a 60% survival, whereas all of the patients in group C (major risk factors) died.

Neonates with mid-small-bowel atresia in group A (no risk factors) or B (minor risk factors) had 82% survival, whereas those in group C (major risk factors) had only a 32% survival .

Survival for atresias involving the distal ileum in neonates in group A (no risk factors) or B (minor risk factors) was 100%; while , for those in group C (major risk factors) survival dropped to 50%.

In our study , newly introduced baseline scoring system of 50 points depend on the estimated risk scores and the P. values for each risk factors . The highest score of six points is related to prematurity and late presentation after the 7th day of life that associated with sepsis . Direct positive correlation was found between the outcome and the increased score digits .

Table 8: Mortality rates of different studies .

Authors	Year Perio	No.of cases	Mortality(%)
Rescorla and Grosfeld ,1985	**1972-1984**	**52**	**13**
Dalla Vecchia *et al* ,1998	**1972-1997**	**128**	**16**
Kumaran etal[6],2002	**1991-1998**	**30**	**0**
Chirdan et al Nigeria 2004	**1998-2002**	**24**	**41.7%**
Thamar H.Stollman	**1971-2004**	**114**	**11**
Ahmed A. Khalaf 2009	**2004 -2007**	**60**	**35**
Our recent study	**2010 -2012**	**64**	**25**

Conclusions

1. The mortality rate of patients with JIA in this recent study was (25%) .
2. Categorization of the patients risk factors according to their estimated risk scores in to: major , minor or no risk factors explained different mortality rates for each category .
3. Preterm was significantly a strong predictor risk factor for mortality with a risk score of 5.4, (P=0.007).
4. The estimated risk score increased with the advancing age of presentation; the risk score of presentation after the 7th day of life was 5.3 (p=0.003).
5. Patients presented with sepsis had a higher risk of mortality as compared to those without sepsis with the risk score of 3 and P<0.05.
6. Jejunal atresia had associated with higher mortality rate, the risk score for jejunal site was 2.8 and P=0.013.
7. Jejunoileal atresia Type III b carried the higher risk (2.5, p=0.017).
8. Anastmosis without plication had much higher estimated risk of 5.4 and P < 0.001.
9. Reoperation was a significant risk factor with an estimated risk score of 2.8, P=0.022.
10. It had been significantly found that mortality rate was directly correlated to the score value that was produced in this study .

Recommendation

We greatly hope to decrease the mortality rate by improving our medical services regarding :

1. Prompt screening for the associated congenital anomalies .
2. Better intensive care facilities , well trained medical personnels & TPN use .

3. Health education of the families and the health care providers in the remote general hospitals and dispensaries regarding clinical presentation of neonatal intestinal obstruction for early referral to pediatric surgery centers .
4. resection of the dilated segments and plication or tapering enteroplasty should be conducted as a protocol for management in all pediatric surgical centers .

References

Ahmed A. Khalaf (2010). Jejunoileal Atresia A study of 60 cases in children welfare teaching hospital .pediatric surgery.

Arnold G. Coran , (2012). Scott Adzick . Jejunoileal atresia and stenosis. Pediatric surgery. Philadelphia 2012 by Saunders company , 7th edition. Chapter 82 : 1059 – 1071 .

Chirdan L.B. A.F. Uba S.D. (2004). Pam Intestinal atresia :management problems in a developing country 2004 .

Dalla Vecchia L ; Grosfeld JL; West KW; Rescorla FJ; Scherer LR; Engum SA(1998). Intestinal atresia and stenosis: a 25-year experience with 277 cases. Arch . Surg . 1998; 133:490-7.

Ekwunife, OH ; Oguejiofor, IC ; Modekwe, VI ; Osuigwe AN(2011). Jejuno-ileal atresia: A 2-year preliminary study on presentation and outcome.pediatric surgery .

Evans CH. (1951). Atresias of the gastrointestinal tract. Int Abstr Surg ;92:1-8.

George Whitfield Holcomb III, J Patrick Murphy; Daniel J. Ostlie (2010): duodenal and intestinal atresia and stenosis. Ashcraft's Pediatric surgery . 5th edition. Philadelphia by Saunders company , chapter 31 : 405 – 415.

James A.O. Neil Jr, Marc I. Rowe, Jay L. Grosfeld. (2006) Jejunoileal tresia. Pediatric surgery. 6th edition. Philadelphia by Mosby Year Book Inc. 2006. Chapter 80:1269-1287

Kumaran N., Shankar KR , Lloyd DAN, Losty PD (2002): Trends in the management and outcome of jejuno-ileal atresia. Eur . J. Pediatr .Surg. 12:163-167.

Lewis Spitz, Arnold G. Coran (2006). Operative pediatric surgery. 6th edition. London 2006 by Hodder Arnold, chapter 43:393-404.

Nixon HH; Tawes R. (1971). Etiology and treatment of small intestinal atresia: analysis of a series of 127 jejunalileal atresias incomparison with 62 duodenal atresias .Surgery; 69:41-51.

Phelps S., Fisher R., Partington A., Dykes E. (1997. Prenatal ultrasonographic diagnosis of gastrointestinal malformations .J . Pediatr. Surg.;32:h44018.

Rescorla FJ, Grosfeld JL. (1985). Intestinal atresia and stenosis: Analysis of survival in120 cases.Surgery; 98(4):668-76.

Sadler. T.W. Langman's Medical Embryology . Part two :chapter 13 : 304 – 308 .

Sathya Prasad Burjonrappa ; Elise Crete; Sarah Bouchard(2011). Comparative outcomes in intestinal atresia: a clinical outcome and pathophysiology analysis.

Shakya et al.(2010). Management of jejunoileal atresias: an experience at eastern Nepal.

Stoll C; Alembik Y; Dott B; Roth MP (1996). Evaluation of prenatal diagnosis of congenital gastrointestinal atresias. Eur J Epidemiol; 12:611±61620.

Tam PK. , Nicholls G. (1999). Implications of antenatal diagnosis of small intestinal atresia in the 1990s . Pediatr. Surg. In ;15 :48 6±48721.

Thamar H.; Stollmana, Ivode Blaauwa(2009) . decreased mortality but increased morbidity in neonates with jejunoileal atresia; a study of 114 cases over a 34-yearperiod.

Vikal C; Shakya , Chandra S; Agrawal1, Pramod Shrestha 2, Prakash Poudel 2, Sudeep Khaniya1, Shailesh Adhikary1 (2010). Management of jejunoileal atresias :an experience at eastern Nepal .pediatric surgery .