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# Is the Incidence of Gastroschisis Increasing Recently?

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## ABSTRACT

**Aims and Objectives:** Gastroschisis is a predominantly full thickness right-sided periumbilical abdominal wall defect. We have observed that the GS birth prevalence has more than doubled over the past few decades. The aim of our study is to point out the increasing incidence of the GS. **Materials and Methods:** We conducted a retrospective study to collect data of the GS, admitted in our institute during the period of 2 years from January 2015 to December 2016. The very high incidence of this congenital anomalies together with the fact that GS is the most common abdominal wall defect requiring neonatal operative intervention and its mortality rate is still very high that prompted us to study its demography, descriptive epidemiology, and quantifying mortality rate and try to analyze factors affecting outcome and cause that might explain this epidemiological pattern. Low- and middle-income countries are often reported to have a higher mortality when compared to patients born in the developed countries. **Results:** A total of 98 patients of the GS were admitted during the period of 2 years. The mortality and survival rate were 26.5% (26) and mortality 73.4% (72) respectively. **Conclusion:** Our data demonstrate that in our institution too, recent trends of increasing prevalence of GS are real. Still, we had a high mortality rate. This study highlights the need for further research to find out the causes of such high incidence rates and to improve our outcomes.

**Key words:** Gastroschisis; Incidence; Mortality

## INTRODUCTION

Maternal age is a key risk factor for the gastroschisis (GS). The incidence among mothers aged under 20 years is 4.71 per 10,000 total births compared with 0.26 per 10,000 total births to mothers aged 30–34 years [1].

GS cases are more likely to be born prematurely and to have poor fetal growth. It is a distressing condition for parents, and some authors highlight the problem that in low-income countries there is an additional challenge that there is poor expectation for survival among medical, nursing, and support staffs. These views are usually conveyed to the parents which may negatively influence health-seeking behavior and may be a deterrent force to seek active management of infants with GS. It requires immediate postnatal surgery, which together with early access to care and a prompt preintervention resuscitation can reach at a good outcome. In developed countries, overall prognosis is good in the majority of infants with an overall postnatal survival rate estimates approximately 90–97% [2].

The aim of our study was mainly to try to explain such a high incidence at our center. We conducted a retrospective review of our data collected from the NICU. Our study was retrospective. We tried to find out the causes of the high incidence retrospectively with data collection. We tried to investigate the causes of the high mortality at our center, to find factors for the improvement in the care and survival rates. It needs further research to reach at the confirmation of the etiological factors of such a high incidence in this geographical area with preclinical and experimental studies on animal models.

## MATERIALS AND METHODS

We conducted a retrospective study to collect data of the GS, admitted in our institute during the period of 2 years from January 2015 to December 2016. We

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collected data including the number of the patients, average age at the operation, male:female ratio, average weight, mothers' history of smoking and drug abuse, average time of the transfer, number of the patients undergoing primary closure (PC) or silo repair, survival rate, and mortality rate. The very high incidence of these congenital anomalies prompted us to study in detail its demography and various factors affecting post-operative outcomes. The study focuses on incidence of this disease in western India.

## RESULTS

A total of 98 patients of the GS were admitted during the period of 2 years (Figure 1). The average age at operation was 1.5 days, male:female ratio 1.5:1, average weight 1.4 kg (Table 1), average time of the transfer >5 h (Table 2), PC in 80 (81.6%) and silo repair (using urobag because preformed silo is not available) in 18 (18.3%) and survival rate - 26.5% (26) and mortality - 73.4% (72). In our study, 10 (10.2%) had atresia, 4 cases had perforation, and 82 (84%) cases had guts matted, edematous and covered with peel. Seventy percent of patients were premature in our study (Table 3). There was a history of smoking in only 10% of cases while there was no history of cocaine use in our study.

## DISCUSSION

Regional registers for congenital anomalies aim to include all data from abortions, fetal loss, and infant deaths, as well as cross-referenced information from pediatric surgical units. Such data sources have consistently shown better and more complete registration of congenital anomalies and have confirmed both an increasing incidence of GS among babies of teenage mothers and an overall increase every year [3].

Recent data from the British Isles Network of Congenital Anomaly Registers confirm the increasing incidence of GS – from 2.5 per 10,000 total births in 1994 to 4.4 per 10,000 in 2004 [4,5]. Among babies of women aged under 20 years, the incidence of GS increased from 8.9 to 24.4 per 10,000 births. Only Li et al. from China presented that the prevalence of the GS is in a decreasing trend [6].

The observed increasing incidence of GS over time seems to be associated consistently with younger maternal age [1]. GS probably does not have a genetic cause because it occurs sporadically, with a relatively low recurrence rate. The most likely cause is early interruption of the fetal omphalomesenteric arterial blood supply. This may be associated with periconceptional tobacco smoking and use of recreational drugs such as alcohol, marijuana, cocaine and use of serotonin uptake inhibitors, oral contraceptives, antivitals, and sexually transmitted infections [7]. While in our study, this has not been supported. The evidence for these associations is, however, only tentative and needs confirmation by carefully controlled cohort or case–control studies [8]. Along with data from regional registers, such studies may lead the way to understand the pathogenesis of this distressing condition and thus preventing it. There is a pressing need to discover the cause of this pandemic trend and thereby eliminate or at least decrease the prevalence of this anomaly, and sociodemographic contexts must be taken into account too to optimize outcomes.

Management involves prompt and appropriate resuscitation, cardiorespiratory support, and initial bowel coverage then reduction of viscera. The latter can be achieved by primary or staged closure. Many infants with GS have a degree of intestinal dysmotility that delays enteral feeding, hence, the requirement for parental nutrition (PN) support that can be a cause of longer hospital stay [9].

The high mortality rate should be put firmly in relationship with geographical and socioeconomical factors in our institution. It is advances in fetal medicine, intensive care, and total parenteral nutrition (TPN) that have resulted in the fall of mortality rates from GS in the developed world from 60% in the 1960s to 4% more recently [9].

In our view, the following factors were the responsible for such a high mortality: Prematurity, fetal growth restriction and low birth weight, lack of antenatal diagnosis, delayed referral with sepsis, delivery and surgical facilities not being under same roof, non-availability of preformed silo, significantly smaller surgeon to patients ratio, poor neonatal intensive care facilities, and lack of enthusiasm of parents to get these babies treated in hospitals.

Sekabira and Hadley previously reported that 91% of cases were born outside their tertiary center in Durban and travelled long distances with resultant hypothermia, dehydration, sepsis, and bowel necrosis [10]. Wright et al. reported similar results in their study [11]. All of our patients were delivered outside our center because gynecological and pediatric surgical departments are not under the single roof. The average time of the transfer was more than 5 h.

Abdur-Rahman et al. noted that despite antenatal ultrasound being available in Ilorin, Nigeria, prenatal diagnosis remained poor with only 1 of the 7 GS cases in their study being detected [12]. In our study, 40% of the patients that were not diagnosed in antenatal ultrasonography. Further, it leads to unplanned management of the GS cases. Antenatally diagnosed cases should be planned in the institution or hospital where pediatric surgery facilities are also available to avoid a delayed referral. Delivery outside a tertiary care cen-

Table 1: Birth weights of patients

| Birth weight of the patients | Number of the patients (%) |
|---|---|
| 1–1.5 kg | 30 (30.61) |
| 1.6–2.0 kg | 40 (40.81) |
| 2.1–2.5 kg | 20 (20.40) |
| >2.6 kg | 08 (08.61) |

Table 2: Time elapsed before transfer

| Duration of the transfer | Number of the patients (%) |
|---|---|
| 1–3 h | 20 (20.40) |
| 4–6 h | 40 (40.81) |
| >7 h | 38 (38.77) |

Table 3: Gestational age

| Gestational age | Number of the patients (%) |
|---|---|
| 32–34 weeks | 20 (20.40) |
| 34–37 weeks | 50 (51.00) |
| >37 weeks | 28 (28.57) |

ter has been associated with increased complications rate. In our study, most of the patients were from the rural area without antenatal diagnosis.

Wright et al. divided countries in the low income countries (LIC), middle income countries (MIC), and high income countries (HIC) [11]. The LIC are included in our study. LIC delegates stated that prenatally diagnosed cases of GS are delivered prematurely [11]. In our study, 70% of patients were between 32 and 36 weeks and birth weight was 1.2–1.8 kg. It is unclear whether this is natural or secondary to medical intervention. Elective preterm delivery has been advocated to reduce bowel exposure to potentially irritant amniotic fluid [13,14]. However, several studies, including a randomized controlled trial and a Cochrane review, have demonstrated that prematurity does not confer survival or functional advantages in infant with GS [15]. Indeed, in the context of scarce neonatal support facilities, premature delivery would almost certainly be disadvantageous. Our institute has a heavy workload (5000 surgeries per year) and working in a condition with fewer surgeons and more patients. Furthermore, we work in a place where all these unfavorable conditions exist which demonstrates that LIC have poor outcome due to lack of medical

Figure 1: Three cases of a gastroschisis admission on a single day

capacity to face this high rate of premature babies delivered without neonatal intensive care facilities.

Whenever PC is not feasible, delegates reported the majority of staged closure in LIC is undertaken using custom silos; preformed silos (PFS) are unavailable or unaffordable in our place. It has been shown that staged closure using PFS reduces the risk of abdominal compartment syndrome and pulmonary barotraumas and improves early renal function [16,17]. Our high rate of the mortality may be due to non-availability of the preformed silo and PC in 80% of the patients. The use of PFS in LICs has potential advantages allowing appropriate resuscitation of the sick infant after transfer in and the possibility of avoiding theater altogether.

Lack of TPN and neonatal intensive care facilities is a problem in sub-Saharan Africa, with reported availability of only 19% and 36%, respectively [11]. The use of TPN has been controversial given the cost implications. Only 19% of LIC surgeons had access to TPN compared with 100% of those in HIC [11]. We consider it an essential part of the package designed to improve outcomes from GS. We are also in favor of using TPN and neonatal intensive care facilities to improve the outcome in cases of GS.

In addition to resuscitation, training for PFS application may be valuable. Dabbas et al. have previously described the use of an inexpensive, easy to construct model for this purpose [18].

A model of an infant with GS was constructed to allow application of a PFS. Each step of the clinical application of a PFS could be simulated. Pediatric surgeons at a regional meeting participated in evaluating the model. This cohort was surveyed with regard to previous clinical experience applying the PFS, invited to apply the silo on the model and then resurveyed with regard to the technique, ease of the application of the PFS on the model, its robustness and potential use as a training tool [18].

We propose the implementation of combined epidemiological research, service delivery training, and resource provision projects to help improve our understanding of GS and outcome. The educational program related to the GS will be helpful for the community.

Training programs that show the great importance of good quality of preintervention resuscitation, adequate intravenous hydration, avoidance of compartment syndrome, and surgical closure's different approaches with subsequent nutritional support are necessary for the best management.

Over the past 5 years, the incidence of cigarette smoking has increased in pregnant teenagers compared with women 20 and over [19]. The clustering of GS in younger mothers and the observed increase in incidence of this malformation over the past 7 years indicates a possibility of an association between cigarette consumption and an interruption of fetal omphalomesenteric arterial blood supply [20].

Insecticides and pesticides used in agriculture based on the history given by parents which belonged to low socioeconomic strata and used these chemicals to save crop in Rajasthan state of India due to low availability of water. This region is also having maximum number of pouch colon cases in world. The atrazine's and nitrates found in these insecticides are responsible for various congenital anomalies. The aldrin, 48 and urea are being used most common insecticides.

Higher concentrations of atrazine in drinking water have been associated with abdominal defects, GS, and other defects [21]. In our view, its need further research to find out association of the GS with the insecticide and pesticide.

Raveenthiran [22] proposed that observations are not superior to hypotheses in generating scientific progress, nor are hypotheses superior to observations. Both are necessary. He explained that the ideal research worker may be one who is equally able to generate hypotheses and to test them experimentally. Most scientists are much better at either one or the other activity [22]. We are also at the one end of the hypothetical association of the GS and planning for future project to prove it with the experiment with the ambitious goal of prevent this pandemic increase of GS prevalence.

## CONCLUSION

We had a large number of cases of the GS during this period at our tertiary center that confirms the significant increase in the prevalence of GS observed and reported by many authors. We had a high mortality rate. We hypothesized the use of some specific pesticides extensively and increasingly used in crop production may have a teratogenic effect, in particular, on younger mothers that are more vulnerable to this exposure through food and drinking water and whose age represents a well know risk's factor for GS. We would not claim so simply that exposure to pesticides is related with high incidence, as there were no such data available from our retrospective study. In our view, it needs further research to find out the cause of such high incidence rate.

## CONSENT

The authors have declared that they have taken consent from patients or their legal guardians regarding publishing of clinical material, in any form, without revealing their identity.

## AUTHORS' CONTRIBUTION

All authors contributed equally in concept, drafting, literature review, and final approval of the manuscript.

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