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SURVIVAL RATE OF PATIENTS WITH CARDIOTHORACIC INJURIES IN ROAD TRAFFIC ACCIDENTS, AND THEIR RELATIONSHIP WITH ISS, GCS AND BLOOD TRANSFUSIONS

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Abstract. Survival Rate of Patients with Cardiothoracic Injuries in Road Traffic Accidents, and their Relationship with ISS, GCS and blood transfusions. Khalid Fahad Almalki, Abdullah Mussad Alharbi, Anthony Morgan, Adel Mohammed Bin Sultan, Saud Ayedh Alajmi, Turke Ali Alajmi, Arif Mohammed Alanazi, Mohammed Ibrahim Alsheddi, Ali Abdulaziz Alanzan, Zayed Fahd Al-Dosari. *Severe thoracic trauma is one of the major causes of injury-related mortality. In the United States, thoracic trauma results in one-fourth of all trauma deaths. Globally, cardiothoracic trauma is also a major contributor to mortality. The most common cardiothoracic injuries include rib fractures, thoracic vertebral fractures, haemothorax, pneumothorax, flail chest, and lung contusions. The purpose of the present study was to determine the survival rate of patients with cardiovascular injuries in road traffic accidents and its relationship with ISS, GCS and blood transfusions at King Khalid Hospital. This study is a useful addition to the literature, as research in this topic is lacking. A total of 189 patients were transported to the hospital with cardiothoracic injuries during the study period. Data was gathered regarding age, gender, nationality, vehicle user type, anatomical region injured, Intensive Care Unit (ICU) admission, Glasgow Coma Scale (GCS), Injury Severity Score (ISS), blood transfusion, treatment and mortality rate. The neurological status was assessed using the GCS score. Injury Severity Scores were calculated to categorize the injury severity. The mean patient age was 31.81 years, with a peak age of between 21–30 years. Males predominated (93.7%) with a male to female ratio of 15:1. Most of the patients were Saudi nationals (61.3%). Overall mortality was 7.9%. Factors that were significantly associated with mortality were head and neck involvement, ICU admission, age (above 60), treatment delivered, and blood transfusions. Cardiothoracic trauma is associated with a high mortality rate, which may depend on the clinical presentation such as GCS, ISS, degree of shock, pattern of injuries, and associated injuries. Immediate management is vital for patients with life-threatening cardiothoracic trauma, as mortality is high if the diagnosis is missed, wrong or left untreated.*

Реферат. Виживання пацієнтів з кардіоторакальними травмами при дорожньо-транспортних пригодах та їх зв'язок з ШТТ, ШКГ та переливанням крові. Халід Фахад Альмалкі, Абдулла Муссад Альхарбі, Ентоні Морган, Адель Мохаммед Бін Султан, Сауд Аєд Альаджмі, Турке Алі Альаджмі, Аріф Мохаммед Альаназі, Мохаммед Ібрагім Альшедді, Алі Абдулазіз Альанзан, Заєд Фахд Аль-Досарі. *Тяжка грудна травма є однією з основних причин смертності, пов'язаної з травмою. У Сполучених Штатах торакальна травма призводить до однієї четвертої частки випадків смертей від травм. У всьому світі кардіоторакальна травма також є основною причиною смертності. Найбільш поширеними кардіоторакальними травмами є переломи ребер, переломи грудного відділу хребта, гемоторакс, пневмоторакс, хитка грудна клітка та*

легеневі контузії. Мета цього дослідження полягала у визначенні показників виживання пацієнтів з серцево-судинними травмами в дорожньо-транспортних пригодах та їх зв'язку з ШТТ, ШКГ та переливанням крові в лікарні короля Халіда. Це дослідження є корисним доповненням до літератури, оскільки недостатньо досліджень з цієї теми. Усього в досліджуваний період до лікарні було перевезено 189 пацієнтів з кардіоторакальними травмами. Зібрано дані щодо віку, статі, національності, типу користувача транспортного засобу, пошкодження анатомічної ділянки, надходження в палату інтенсивної терапії (ПІТ), шкали коми Глазго (ШКГ), показника шкали тяжкості травми (ШТТ), переливання крові, рівня лікування та смертності. Неврологічний статус визначали за допомогою оцінки ШКГ. Оцінка тяжкості травми була розрахована для класифікації тяжкості травми. Середній вік пацієнтів становив 31,81 року, а більше всього пацієнтів віком від 21 до 30 років. Чоловіки переважали (93,7%), а співвідношення чоловіків і жінок становило 15:1. Більшість пацієнтів були громадянами Саудівської Аравії (61,3%). Загальна смертність становила 7,9%. Факторами, які суттєво асоціювалися зі смертю, були ураження голови та шиї, надходження в ПІТ, віку (старше 60 років), лікування, а також переливання крові. Кардіоторакальна травма асоціюється з високим рівнем смертності, що може залежати від клінічної картини, такої як ШКГ, ШТТ, ступінь шоку, тип травми та супутніх з ними травм. Негайне лікування є життєво важливим для пацієнтів, які страждають на серцево-торакальні травми, які загрожують життю, оскільки смертність є високою, якщо діагноз не поставлений, неправильний або пацієнт не отримував лікування.

Cardiothoracic trauma, either blunt or penetrating in nature, causes significant mortality and morbidity, especially in developing countries [1, 2].Thoracic trauma is a leading cause of death in 25-50% of cases that have multiple traumas [3]. In United States, thoracic trauma results in one-fourth of all trauma deaths [4]. Isolated penetrating chest injury accounts for mortality in less than 01% of cases; however, the mortality rate increase to 20% due to hypoxia and hypovolemia if cardiac injury coexists [3]. Road traffic accidents (RTA) account for 60-70% of blunt chest trauma, a major cause of hospitalization [1, 5]. Globally, cardiothoracic trauma is also a major contributor to mortality. The most common cardiothoracic injuries include rib fractures, thoracic vertebral fractures, haemothorax, pneumothorax, flail chest, and lung contusions [6]. Cardiac tamponade and injury to the great vessels may also occur, but these are uncommon.

The most important issue for patients with cardiothoracic injuries is to prevent a deadly outcome, because such injuries can result in fatal consequences immediately after the trauma has occurred. The conditions, which require immediate emergency management for patients with cardiothoracic trauma include airway obstruction, massive haemothorax, tension pneumothorax, open pneumothorax, flail segment and cardiac tamponade, cardiac dysfunction, and injury to large intra-thoracic vessels. Death is imminent if these life-threatening conditions are missed or left untreated. Additionally, co-morbid conditions can even worsen the outcome [7]. Therefore, it is critical to evaluate and manage cardiothoracic trauma on an emergency basis, as accuracy of diagnosis is often at risk for patients with these conditions. It has been reported that prompt evaluation and improvement in diagnostic accuracy substantially reduces the mortality rate [8]. The management of chest trauma should be dealt with according to the severity of the injury. Life-threatening conditions may require urgent surgical interventions, such as removal of a foreign body from the airway, chest tube insertion, and fluid or blood resuscitation in hypovloemic shock.

Severe thoracic trauma is one of the major causes of injury-related mortality. Several scoring systems have been established to determine trauma severity. Cardiothoracic injuries associated with RTAs can be evaluated based on the type of injury, the trauma and injury severity score (TRISS) or the Injury Severity Score (ISS), the Glasgow Coma Scale (GCS), and required blood transfusion. ISS was developed for patients with multiple traumas, in order to assess trauma severity in terms of morbidity and mortality [9]. It is an established and validated anatomical system, which records an overall trauma injury score. Although ISS has some mathematical, administrative and clinical limitations, it is nevertheless regarded as a “Gold Standard” to measure the severity of trauma [10]. The retrospective study of medical records conducted by Moon et al. [11] validates TRISS for the assessment of thoracic trauma severity. Studies have reported a high ISS in patients with cardiothoracic injuries [12]. However, some authors have questioned the validity of ISS for assessing cardiothoracic injuries, as it is not specific for isolated thoracic trauma, and severity may be underestimated [13].

Being simple and reliable, GCS is one of the most common tools used for the assessment and monitoring of a patient’s level of consciousness. Studies have reported a lower GCS in patients with cardiothoracic injuries [12]. Patients with a GCS of 8 or less indicate severe thoracic trauma. A similar prospective study was carried out over three years to determine the factors affecting mortality in patients with chest trauma in United Arab Emirates [14]. They reported that GCS significantly affects the

survival rate in the patients with chest trauma. GCS has some limitations e.g. inter-observer reliability, time passed since the injury occurred and confounding factors [3], which may affect the outcome. Similarly, in another study, GCS was reported as an independent prognostic predictor of mortality in patients with thoracic trauma [15].

Blood transfusion is a life-saving trauma service, especially when massive blood loss and hemorrhagic shock occur in patients with RTA. Therefore, patients with massive blood loss or shock should be managed on a priority basis with adequate amounts of intravenous fluids and blood transfusion. Although blood transfusion is a life-saving critical trauma service, it can nevertheless cause a large number of complications, such as electrolyte imbalance (hypocalcemia, hyperkalemia and hypomagnesemia), disturbed acid–base balance, and hypothermia. The purpose of the present study was to determine the survival rate of patients with cardiovascular injuries due to road traffic accidents, and the relationship of these injuries with ISS, GCS and blood transfusions at King Khalid Hospital. This study is a useful addition to the literature, as research in this topic is lacking.

MATERIALS AND METHODS

A hospital-based cross-sectional retrospective study was carried out at King Khalid Hospital and Prince Sultan Center for Health Services (KKH & PSCHS) from January 01, 2012 to December 31, 2017, to assess the survival rate of patients with cardiothoracic injuries resulting from road traffic accidents (RTA), and correlations with the Glasgow Coma Scale (GCS), the Injury Severity Score (ISS) and blood transfusions. The King Khalid Hospital and Prince Sultan Center for Health Services (KKH & PSCHS) is a three hundred and fifty (350) bed tertiary care referral hospital that serves the people of Al Kharj and neighboring towns. The hospital receives most of the trauma in the city and its outskirts. All victims of road traffic accidents (RTA) with cardiothoracic injuries, admitted to the Emergency Department of the King Khalid Hospital during the study period were recruited in the study.

During the study period, 189 patients were transported to the hospital with cardiothoracic injuries. All of these patients were involved in a traffic accident and were admitted to the emergency department. For this study, the probability sampling technique was used. All patients who met the inclusion criteria during the study period were consecutively recruited in the study until the sample size was reached.

The study subjects were selected based on the following inclusion criteria: 1 – Patients with cardiothoracic injuries. 2- Patients with injuries caused by a road traffic accident (RTA). 3- Patients with a hospital stay of more than 24 hours during the period of the study. The exclusion criteria included: 1 – Patients who did not sustain cardiothoracic injuries. 2 – Patients with injuries not caused by a traffic accident. 3 – Patients with a hospital stay of less than 24 hours. 4 – Patients who had incomplete medical records.

All patients who met the inclusion criteria were approached for consent. A pretested, well-structured questionnaire was used for data collection. A face-to-face interview was conducted by the researcher using a questionnaire in the patient's language. Closed-ended questions were asked. All the other patient data were obtained by scrutinizing the database at the medical records department of the hospital. Medical files were retrospectively reviewed during the study period in order to study patient characteristics.

Data was retrieved regarding age, gender, nationality, vehicle user type, anatomical region injured, Intensive Care Unit (ICU) admission, Glasgow Coma Scale (GCS), Injury Severity Score (ISS), blood transfusion, treatment and mortality rate. The neurological status was assessed using the GCS score. Injury Severity Scores were calculated to categorize injury severity. All the cases were subdivided into the groups according to the injured body area: upper and lower limbs, head and neck, chest, abdominal, pelvic, spine and multiple injuries.

The quality of data was maintained by incorporating only the complete data of the subjects within the study period. The collected data was checked at different levels for completeness and consistency by the principal researcher at the end of each day. Whenever an error was found at any level, it was corrected as needed.

The Statistical Package for the Social Sciences (SPSS) computer software, version 20, was used for entry, compilation and analysis of data for statistical significance. The quantitative variables such as age and its categories were presented by calculating mean and standard deviation. For qualitative variables such as gender, nationality, and type of injury, frequency and percentage distribution tables were generated. The chi-square test and z-test were applied for qualitative and quantitative variables respectively. Probability (P-values) less than 0.05 were taken as statistically significant. The relationship between the severity of injury and ISS, GCS and blood transfusions were studied. The significance of relationship was also assessed. The data was presented by using statements, figure and tables. The Mann-Whitney U test was applied to assess the

comparison. Effect modifiers such as age, gender and the severity of the accident were controlled through stratification. Variables analyzed included chest involvement, neck injury, age, gender, injury severity, neurological involvement, surgical procedures and transfusion. Data was reported as frequency and mean ± standard deviation.

Formal approval was given by the ethics committee and research department of the King Khalid Hospital and Prince Sultan Center for Health Services (KKH & PSCHS). The utmost effort was made to ensure that all the ethical standards of research were applied. All study participants were briefed with the research purpose and the confidentiality of the information collected. A written consent was provided by each participant. The participants were advised that they had the right not to participate, and could exclude themselves from the study at any point in time during the study if they felt uncomfortable.

RESULTS AND DISCUSSION

Total of 189 patients were recruited for the study based on inclusion criteria. The mean patient age was 31.81 years with a peak age between 21-30 years. Males predominated (93.7%) with a male to female ratio of 15:1. Most of the patients were Saudi nationals (61.3%). The socio-demographic characteristics are shown in Table 1.

Table 1

Socio-demographic characteristics of the patients along with study variables

		Frequency	Percent
Age Category	**0-10**	**9**	**4.8%**
	11-20	**33**	**17.5%**
	21-30	**63**	**33.3%**
	31-40	**48**	**25.4%**
	41-50	**9**	**4.8%**
	51-60	**15**	**7.9%**
	above 60	**12**	**6.3%**
Nationality	**Saudi**	**114**	**61.3%**
	Non-Saudi	**72**	**38.7%**
Gender	**Female**	**12**	**6.3 %**
	Male	**177**	**93.7%**
Mortality	**Alive**	**174**	**92.1%**
	Dead	**15**	**7.9%**
Rib Fractures	**Yes**	**123**	**65.1%**
	No	**66**	**34.9%**
Operation	**Yes**	**99**	**52.4%**
	No	**90**	**47.6%**
Icu Admission	**Yes**	**42**	**22.2%**
	No	**147**	**77.8%**
Shock degree	**1st degree**	**63**	**33.3%**
	2nd degree	**21**	**11.1%**
	3rd degree	**6**	**3.2%**
	No shock	**99**	**52.4%**
Accident Cause	**Car**	**183**	**96.8%**
	Motorbike	**6**	**3.2%**

Only road traffic accidents were included. Of these, cars were involved in 96.8% of accidents. Most of these patients (92.1%) survived and recovered after their accident. All patients were reported to have non-penetrating chest injuries. The pattern of chest injuries was predominated by single or multiple rib fractures (65.1%), followed by lung contusions (46%). A simple or tension pneumothorax was present in 19% of patients, and pleural effusion was present in 23.8% of patients. Hemithorax developed in 22.2% of the cases. Chest laceration and chest drain was reported in 41.3% and 42.9% cases respectively. Of the total rib fractures reported (65.1%), 31.7% developed two fractures and 33.3% developed multi-fractures. An associated neck injury was also reported in more than half of these patients (Fig. 1)

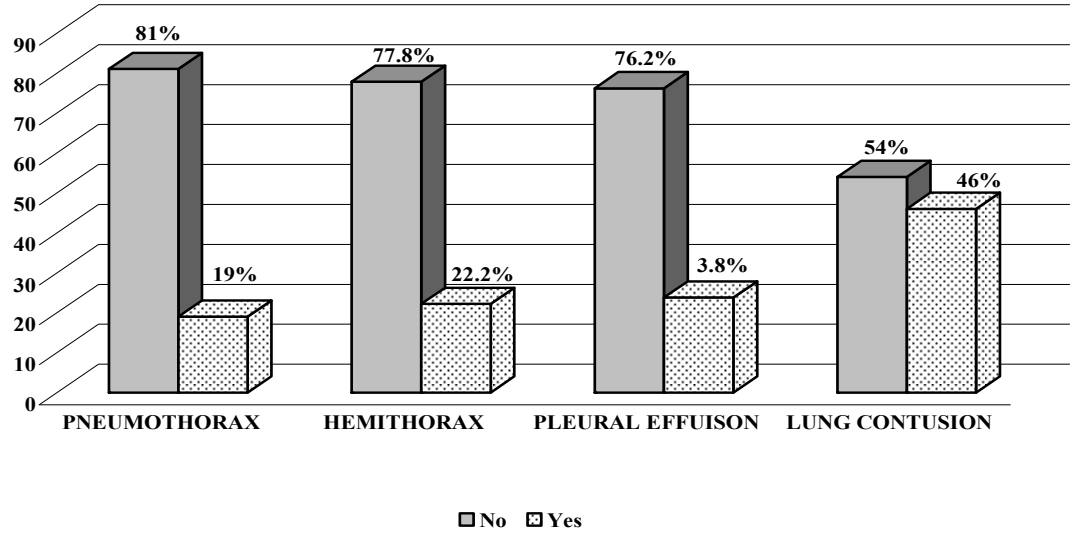

Fig. 1. Pattern of chest injuries

For treatment, 90 (48.6%) victims underwent conservative therapy. Overall, 99 (52.4%) victims required surgery, out of which laparotomy (32.4%) predominated, followed by suturing (16.2%), splenectomy (12.7%) and rib cast (2.7%) (Fig. 2).

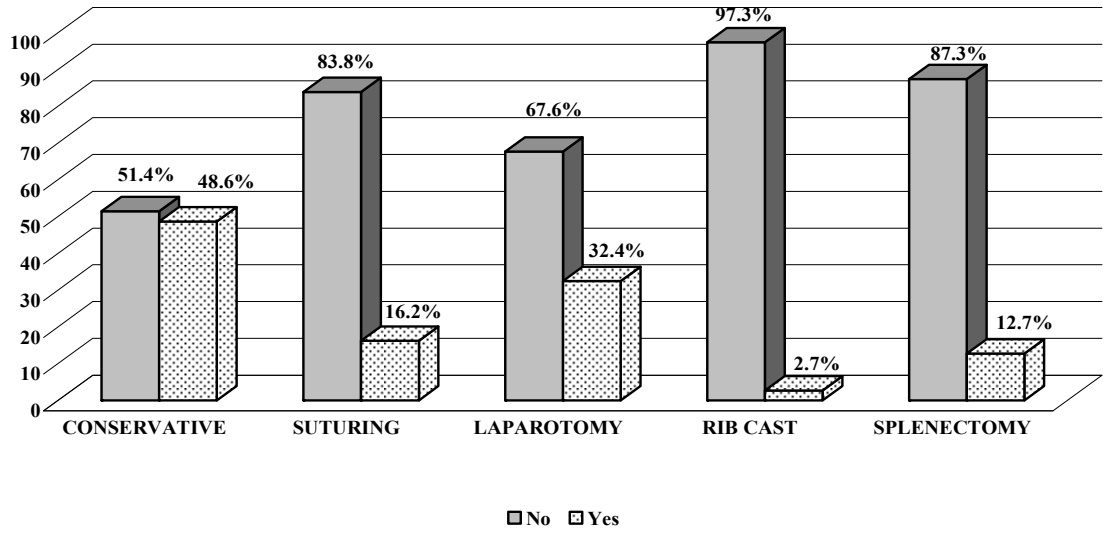

Fig. 2. Management of cardiothoracic injured patients

More than half (52.4%) of the patients did not develop shock and were stable. Of those remaining, 33.3% developed 1st degree shock, 11.1% developed 2nd degree shock, and only 3.2% were reported to have developed 3rd degree shock, which requires immediate blood transfusion. About one-third (36.5%) of patients received a blood transfusion (Table 2). A two-way cross-tabulation was performed to compare rate of mortality with the degree of shock in RTA victims. It was inferred from the two-way cross-tabulation that the degree of shock was low in surviving patients (56.9% of survivors had no shock). Victims with circulatory shock and multiple injuries were identified as having a higher rate of mortality. However, the Pearson Chi Square test could not be performed because of inadequate conditions.

Table 2

Relationship between degree of shock and mortality rate mortality * shock_degree Cross-tabulation

			Shock_Degree				Total
			no shock	1st degree	2nd degree	3rd degree	
Mortality	Alive	Count	99	57	15	3	174
		% of Mortality	56.9%	32.8%	8.6%	1.7%	100.0%
	Dead	Count	0	6	6	3	15
		% of Mortality	0.0%	40.0%	40.0%	20.0%	100.0%
Total		Count	99	63	21	6	189
		% of Mortality	52.4%	33.3%	11.1%	3.2%	100.0%

A total of 22.2% of patients were admitted to the ICU from the accident and emergency department. The mean (SD) blood transfusion was 2.22 (1.29) units. The mean (SD) ICU and hospital stay was 7.71 (7.43) days and 10.37 (12.61) days respectively (Table 1).

Overall mortality was 7.9%. Factors that were significantly associated with mortality were head and neck involvement, ICU admission, age (above 60), treatment delivered and blood transfusion. Mortality was also influenced by injury severity score ISS ($p<0.001$) and the Glasgow Coma Score GCS ($p<0.000$). A total of 92.1% of patients had mean GCS scores of 13.81, and the remaining 7.9% had low mean GCS scores of 4.60, which was associated with poor prognosis and high mortality. So, the mean GCS of surviving patients was significantly higher than those who died. In other words, deceased patients had a lower GCS than surviving patients.

The mean ISS was 24.75 for surviving patients. The mean ISS of deceased patients was 40.40, which was higher than that of patients who survived. A high ISS was associated with severe injury and high mortality. According to the relation between ISS and mortality, all deaths were among patients with severe injuries; all other cases survived. Patients with higher a GCS score had a lower ISS. The probability of survival increased with increasing GCS scores (Table 3).

Road traffic accidents (RTA) are the most frequent cause of cardiothoracic trauma that contributes significantly to morbidity and mortality. In the present study, all patients encountered RTA-related non-penetrating thoracic trauma, with survival and mortality rates of 92.1% and 7.9% respectively. Multiple rib fractures, lung contusions, chest lacerations, pleural effusion, hemothorax and simple or tension pneuomothrax were the most common injuries encountered. More than half of the patients had associated neck injuries, and similar number of patients required operation. The study revealed that higher mortality was associated with a high injury severity score (ISS) and a low Glasgow coma scale (GCS). About one-third of the patients developed first-degree shock, while third-degree shock required immediate blood transfusion.

The present study reports a higher mortality rate (9.1%) in the region. The reason behind the high mortality may be attributed to non-penetrating thoracic trauma, higher ISS, lower GCS and the degree of shock reported in all deceased patients. In contrast, previous studies have reported lower mortality rates with cardiothoracic injuries.

A retrospective study that included 378 RTA patients with chest trauma conducted at King Khalid Hospital reported rib fracture in 64.4% patients as the predominant type of chest injury [6]. Other cardiothoracic injuries were injury to thoracic vertebrae (41.5%), haemothorax (26.5%), simple pneumothorax (22.8%), lung contusion (20.1%), sternal fracture (3.4%), flail segment (2.6%) and cardiac tamponade (0.5%). The study reported a mortality rate of 4.8%. In another study, Lien et al. [16] retrospectively studied the data of 18,856 patients who were admitted to hospital in Taiwan with cardiothoracic injuries after a traffic accident. They reported 24-hour mortality in 2.4% of cases, which comprised of about 55% of all 30-day mortalities. Alam El-Din et al. [17] prospectively studied 100 patients with chest trauma at Menoufia University Hospital in Egypt. They reported that 72% of patients had blunt trauma, and 28% patients had penetrating trauma. The most common cause of blunt trauma was motor vehicle accidents (MVA), while the most common cause of penetrating trauma was stab wounds. They reported a mortality rate of 6.9% among patients with blunt trauma, and no mortality among patients with penetrating trauma.

Table 3

Relationship of mortality with chest injury and ISS

		Frequency	Mean	Std. Deviation
ISS	**Alive**	**174**	**24.75**	**12.419**
	Dead	**15**	**40.40**	**19.316**
p value	**0.001***			
Relation between mortality in chest injury and GCS				
Neck Injury	**Alive**	**174**	**13.81**	**1.930**
	Dead	**15**	**4.60**	**0.828**
p value	**0.000***			

The difference in the mortality rate of the present study and previous studies is due to various reasons. Firstly, in the present study, there were associated head and neck injuries in more than half of the patients, and head and neck injuries are associated with increased mortality. Ekpe and Eyo [18] studied determinants of mortality in patients with chest trauma. They reported a 5.4% mortality rate, and demonstrated that chest trauma with associated head trauma contributes significantly to mortality. El-Menyar, et al. [19] studied 5118 RTA-related traumatic injuries in Qatar, reporting a 15% mortality rate. They reported that head injury was significantly associated with ISS and abbreviated injury score (AIS).

Secondly, all patients had encountered blunt trauma, which leads to a longer hospital stay and a high mortality rate. As mentioned earlier, Alam El-Din [17] found that traffic accidents cause higher rates of blunt trauma to the chest, with higher mortality, as compared to penetrating trauma. Hence, blunt trauma in every patient might have led to higher mortality. Thirdly, higher ISS and lower GCS results in a low survival rate and a high mortality rate. Moon et al. [11] conducted a retrospective study on medical records of 228 patients with severe thoracic trauma, in order to assess mortality using TRISS and TTSS. They reported that a higher TRISS in patients with thoracic trauma contributes to mortality. Similarly, a lower GCS at presentation is associated with higher mortality. Manay et al. [20] conducted a prospective study that included 139 patients with chest trauma. They reported that a GCS higher than 8 in the patients with blunt chest trauma was significantly associated with an increased mortality rate ($p = 0.022$). In the present study, the patients who died had a mean GCS of 4.6, as compared to 13.81 in those who survived. Similarly, massive blood loss is also associated with a higher mortality rate. Although the present study reveals that circulatory shock is associated with increased mortality, statistical evidence could not be presented due to inadequate conditions. In order to determine mortality, Veysi et al. [21] conducted a prospective study that included 1,164 patients with multiple traumas, with at least one chest injury. They divided the patients into six groups of AIS, depending on the number of chest

injuries. Their study revealed no significant effect of blood transfusions in all six groups of AIS.Fourthly, the present study included 38.7% non-Saudi patients, contributing more than one-third population of the study. It may affect the actual results in Saudi population as non-Saudi patients or drivers might have problem while following the traffic rules in KSA. However, this parameter should be reevaluated in further studies.

CONCLUSION

In the present study, 48.6% of victims underwent conservative therapy, while 52.4% required surgery, i.e. chest intubation (42.9%) and laparotomy (32.4%), including suturing, splenectomy and rib cast. Okugbo et al. [22] conducted a prospective study that included 73 patients with chest trauma. They reported that tube thoracostomy was required in 51% of cases and laparotomy in 13.7% of cases. They reported mortality in 2.7% of cases due to open pneumo-hemothorax and massive blood loss. In conclusion, cardiothoracic trauma is associated with a high mortality rate, which may depend on the clinical presentation such as GCS, ISS, degree of shock, pattern of injuries and associated injuries. Immediate intervention is vital for patients with life-threatening cardiothoracic trauma, as mortality is high if the diagnosis is missed, wrong or left untreated.

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МЕДИКАМЕНТОЗНАЯ ТЕРАПИЯ ПАЦИЕНТОВ С ЛЕГОЧНОЙ АРТЕРИАЛЬНОЙ ГИПЕРТЕНЗИЕЙ НА ФОНЕ СИСТЕМНОЙ СКЛЕРОДЕРМИИ (часть 1)

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Ключевые слова: *легочная артериальная гипертензия, системная склеродермия, медикаментозная терапия легочной гипертензии*
Ключові слова: *легенева артеріальна гіпертензія, системна склеродермія, медикаментозне лікування легеневої гіпертензії*
Key words: *pulmonary arterial hypertension, systemic scleroderma, pharmacological treatment of pulmonary hypertension*

Реферат. Медикаментозна терапія пацієнтів з легеневою артеріальною гіпертензією на тлі системної склеродермії (частина 1). Єгудіна Є.Д., Калашникова О.С. *Легенева артеріальна гіпертензія (ЛАГ) − тяжке захворювання, зумовлене ураженням судин легеневого мікроциркуляторного русла, що призводить до підвищення в них тиску, збільшення легеневого судинного опору, правошлуночкової серцевої недостатності та смерті. ЛАГ належить до актуальних проблем сучасної медицини через низьке виживання, швидку інвалідизацію пацієнтів та велику вартість лікування. ЛАГ є одним з основних чинників смертності при системній склеродермії (ССД). ЛАГ, асоційована зі склеродермією (ССД-ЛАГ), є унікальним фенотипом, який поєднує ССД та ЛАГ, патогенетичні механізми котрого модифікують клінічну картину цих станів. Сучасна діагностика та лікування ЛАГ значно впливає на показник виживання, однак раннє виявлення ЛАГ все одно викликає труднощі при ССД через декілька факторів. Перш за все, це обмеження сучасних діагностичних методів скринінгу та поліорганність ураження при ССД. Порівняно з іншими підгрупами ЛАГ, пацієнти з ССД-ЛАГ погано реагують на звичайні форми терапії ЛАГ. ССД-ЛАГ та ідіопатична легенева артеріальна гіпертензія (ІЛАГ) належать до І групи за класифікацією легеневої гіпертензії та відповідно до сучасного уявлення мають схожий патогенез та клінічну картину, однак клінічні відмінності відповіді на терапію демонструють, що в патогенетичних побудовах ССД-ЛАГ можуть брати участь різні механізми. Недавні результати*