Journal of Pediatric Surgery 50 (2015) 1441–1456

Contents lists available at ScienceDirect

Journal of Pediatric Surgery

journal homepage: www.elsevier.com/locate/jpedsurg

Original Articles

A cross-sectional nationwide survey on esophageal atresia and tracheoesophageal fistula

A. Pini Prato [a,*], M. Carlucci [a], P. Bagolan [b], P.G. Gamba [c], M. Bernardi [d], E. Leva [e], G. Paradies [f], C. Manzoni [g], B. Noccioli [h], A. Tramontano [i], V. Jasonni [a,j], F. Vaccarella [k], S. De Pascale [l], D. Alberti [m], G. Riccipetitoni [n], D. Falchetti [o], F. Caccia [p], G. Pelizzo [q], J. Schleef [r], M. Lima [s], P. Andriolo [t], A. Franchella [u], A. Cacciari [v], F. Caravaggi [w], S. Federici [x], M. Andermarcher [y], G. Perrino [z], D. Codrich [aa], F.S. Camoglio [ab], F.S. Chiarenza [ac], A. Martino [ad], A. Appignani [ae], V. Briganti [af], S. Caterino [ag], D. Cozzi [ah], M. Messina [ai], A. Rizzo [aj], L. Liotta [ak], D. Salerno [al], M.G.R. Aceti [am], F. Bartoli [an], C. Romeo [ap], C. Esposito [aq], P.L. Lelli Chiesa [ar], E. Clemente [as], L. Mascia [at], S. Cacciaguerra [au], V. Di Benedetto [av], S. Licciardi [aw], E. De Grazia [ax], M. Ubertazzi [ay], G. Piazza [az], G. Mattioli [aj], F. Rossi [ar], M. Nobili [an]

[a] *Istituto Giannina Gaslini, Genova, Italy*
[b] *Ospedale Pediatrico Bambino Gesù, Roma, Italy*
[c] *Azienda Ospedaliero-Universitaria, Padova, Italy*
[d] *Azienda Ospedaliera della Provincia di Lecco, Merate, Italy*
[e] *Ospedale Maggiore Policlinico Magiagalli, Milano, Italy*
[f] *Ospedale Giovanni XXIII, Bari, Italy*
[g] *Policlinico Gemelli, Roma, Italy*
[h] *Ospedale Pediatrico Meyer, Firenze, Italy*
[i] *Azienda Ospedaliera Pediatrica Santobono Pausilipon, Napoli, Italy*
[j] *Università degli Studi di Genova, Genova, Italy*
[k] *ASN SS: Antonio e Biagio e Cesare Arrigo, Alessandria*
[l] *Ospedali Riuniti, Bergamo*
[m] *Spedali Civili, Brescia*
[n] *Ospedale dei Bambini V. Buzzi, Milano*
[o] *Ospedale Niguarda Ca' Granda, Milano*
[p] *Ospedale San Carlo Borromeo, Milano*
[q] *Policlinico San Matteo, Pavia*
[r] *Ospedale Infantile Regina Margherita, Torino*
[s] *Ospedale Sant'Orsola Malpighi, Bologna*
[t] *Ospedale di Bolzano*
[u] *Archispedale Sant'Anna, Ferrara*
[v] *Ospedale Policlinico, Modena*
[w] *Azienda Ospedalier-Universitaria, Parma*
[x] *Ospedale Infermi, Rimini*
[y] *Ospedale Santa Chiara, Trento*
[z] *Ospedale Regionale, Treviso*
[aa] *Ospedale Infantile Burlo Garofalo, Trieste*
[ab] *Policlinico Borgo Roma, Verona*
[ac] *Ospedale San Bortolo, Vicenza*
[ad] *Ospedale Salesi, Ancona*
[ae] *Ospedale Santa Maria della Misericordia, Perugia*
[af] *Ospedale San Camillo Forlanini, Roma*
[ag] *Ospedale Policlinico Sant'Andrea, Roma*
[ah] *Policlinico Umberto I, Roma*
[ai] *Ospedale Policlinico Santa Maria alle Scotte, Siena*
[aj] *Ospedale Policlinico, Bari*
[ak] *Ospedale Francesco Ferrari, Casarano*
[al] *Azienda Ospedaliera Pugliese-Ciaccio, Catanzaro*
[am] *Azienda Ospedaliera di Cosenza, Cosenza*
[an] *Azienda Ospedaliera Universitaria-Ospedali Riuniti, Foggia*
[ap] *Azienda Ospedaliero-Universitaria G. Martino, Messina*
[aq] *Policlinico Universitario Federico II, Napoli*
[ar] *Ospedale Maggiore della carità, Novara*

* Corresponding author at: Istituto Giannina Gaslini, Largo G. Gaslini, 5, Genova, Italy. Tel.: +39 0105636217; fax: +39 0103075092.
E-mail address: a.piniprato@gmail.com (A. Pini Prato).

http://dx.doi.org/10.1016/j.jpedsurg.2015.01.004
0022-3468/

[as] Azienda Ospedaliera Universitaria S. Giovanni di Dio e Ruggi d'Aragona, Salerno
[at] Ospedale SS. Trinita', Cagliari
[au] Ospedale Garibaldi, Catania
[av] Ospedale Vittorio Emanuele, Catania
[aw] Presdio Ospedaliero CTO, Iglesias
[ax] Azienda Ospedaliero-Universitaria Policlinico P. Giaccone
[ay] Azienda Ospedaliero-Universitaria, Sassari
[az] Ospedale Sant'Antonio Abate, Trapani
[ar] Ospedale Maggiore della carità, Novara

ARTICLE INFO

Article history:
Received 28 October 2014
Received in revised form 7 January 2015
Accepted 9 January 2015

Key words:
Esophageal atresia
Diagnosis
Management
Mortality
Nationwide survey
Cross-sectional

ABSTRACT

Background: Our study aims at disclosing epidemiology and most relevant clinical features of esophageal atresia (EA) pointing to a model of multicentre collaboration.
Methods: A detailed questionnaire was sent to all Italian Units of pediatric surgery in order to collect data of patients born with EA between January and December 2012. The results were crosschecked by matching date and place of birth of the patients with those of diagnosis-related group provided by the Italian Ministry of Health (MOH).
Results: A total of 146 questionnaires were returned plus a further 32 patients reported in the MOH database. Basing on a total of 178 patients with EA born in Italy in 2012, the incidence of EA was calculated in 3.33 per 10,000 live births. Antenatal diagnosis was suspected in 29.5% patients. 55.5% showed associated anomalies. The most common type of EA was Gross type C (89%). Postoperative complications occurred in 37% of type C EA and 100% of type A EA. A 9.5% mortality rate was reported.
Conclusions: This is the first Italian cross-sectional nationwide survey on EA. We can now develop shared guidelines and provide more reliable prognostic expectations for our patients.

Esophageal atresia (EA) is a rare disease and represents the most frequent congenital anomaly of the esophagus. The etiology is still unknown but environmental and/or genetic factors have been suggested [1–3]. The epidemiology of EA has been shown to vary in reported series with prevalence raging from 1:2500 to 1:4500 live births [1,2,4–11]. The most frequently encountered variant (75–90% of cases) is EA with distal TEF, type C according to Gross classification [1,7,10–13]. More than 50% of EA patients have associated anomalies involving organs and systems [2,5,7,8,11,12,14]. Advances in surgical techniques and in intensive neonatal care improved survival over the past decades, with a rate now approaching 90% also in infants with severe associated anomalies and 100% in those without [2,7,8,12,15–18]. The absence of an international data collection system makes providing a reliable EA epidemiology very difficult, which is of outmost importance in order to identify risk factors, provide better prognostic expectations and educate families. At present, most of reports on EA are based on few single institution retrospective or population-based studies, focused on descriptive epidemiology and survival rates. Inspired by other national and international cohort studies [1,5,7,11], our study addressed the epidemiology and most relevant clinical features of EA in Italy, focusing on a model of multicentre collaboration similar to the previously reported by Sfeir and co-workers in France, and Burge and colleagues in the United Kingdom [5,11]. This study aims at providing reliable epidemiological data for physicians dealing with this rare congenital disease in Italy as well as abroad. Furthermore, we will provide detailed information regarding clinical features, short term outcome and survival that will turn extremely useful to a reliable prenatal and/or postnatal counselling. Finally, the results of our study will hopefully help in implementing nationally shared guidelines to improve the overall outcome of our patients.

1. Materials and methods

The Italian Society of Pediatric Surgery (ISPS) Directorate implemented this prospective observational cross-sectional study project during the 42nd national Congress that was held in Padua in September 2011. Resorting to the national Ministry of Health (MOH) database cross-matched with the ISPS database we could identify and enrol a total of 52 Units of Pediatric Surgery dealing with newborn surgery in Italy. A questionnaire was sent to each responsible physician who was asked to send back the completed questionnaire immediately after patients' discharge from the hospital (the list of responsible physician in each Unit is available in Appendix 1). The questionnaire was implemented by a committee of pediatric surgeons (experts from the ISPS directorate) and addressed various issues (63 to 69 items based on type of EA) including demography (5 items), family history (3 items), pregnancy (7 items), perinatal period (4 items), associated anomalies (9 items), clinical features and perioperative management (12 items), surgical details according to type of EA (10 to 16 items), postoperative information (4 items), morbidity and early mortality (within 30 days of life) (9 items) (Appendix 2).

1.1. Definitions

EA was classified according to Gross classification [19] and risk groups were defined according to Spitz classification [20]. Surgical details, complications and short term outcome were addressed separately for type A/B and type C/D EA given the similarity of those EA types. Similarly, type 5 EA, not requiring esophageal anastomosis, underwent specific considerations. VACTERL association was defined when at least 3 of the following congenital anomalies were also present: vertebral, anorectal, cardiac, renal, urinary, and limb abnormalities.

1.2. Inclusion/exclusion criteria

The questionnaire was sent in November 2011 to all Italian Units of pediatric surgery. Patient's inclusion criteria were as follows: 1) neonatal confirmed diagnosis of EA/TEF; 2) date of birth between the 1st of January and the 31st of December 2012. Exclusion criteria were as follows: 1) stillborn with EA/TEF, and 2) voluntary pregnancy termination owing to EA/TEF suspicion. Deadline for collecting questionnaires was set on the 30th of June 2013 to allow the inclusion of late responders. The questionnaire included all data collected by the surgeon in charge of the patient at first discharge from the hospital, excluding those concerning esophageal strictures that were collected throughout the