The rise and decline of human hydatid disease in Portugal: historical and epidemiological analysis
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Abstract
The author analyses here the official statistics and his own 30-year casuistry about human hydatid disease, for the purpose of clarifying the fallacious statement that Portugal, in general, and the Alentejo, in particular, is hyper endemic in this parasitical zoonosis. Thus, the occurrence of hydatid disease is analyzed at several levels: Country, Great Regions, Alentejo, Evora district, and the counties of this district.

The final conclusion is that the majority of the Country is, in general, sine endemic in human hydatid disease, the only occurrences being sporadic cases that are, more often than not, the result of old infections. As far as the Alentejo is concerned – in the past the largest endemic region of Portugal – there are counties in the districts of Portalegre and Beja that are sine endemic, and some hypo-endemic; only the district of Evora has a few counties that are meso-endemic and only one that is hyperendemic.

Key-words: hydatid disease, epidemiology, hydatid disease incidence, Portugal, Alentejo.

INTRODUCTION
In the past, echinococcosis/hydatidosis began to increase in clinical and epidemiological importance in south Europe. However, good control programs have managed to significantly control or even eradicate this parasitic zoonosis in the main countries affected. In Portugal, there is an almost total lack of information with regard to echinococcosis, i.e. on the definitive infection of the host, the dog (the only global study) is clearly outdated as it was conducted around forty years ago. With regard to animal hydatidosis, there was, until 1968, a large amount of official information on inspections in slaughterhouses, but nowadays, if such statistics exist, they are incomplete or inaccessible. In relation to human hydatidosis, the official data are clearly unsatisfactory; however, in conjunction with the case studies from Evora, the district with the highest incidence rates in the country, it is possible to build up a fairly accurate concept of the evolution of this parasitosis in Portugal, both temporally and spatially.

It turns out that for reasons that are not entirely plausible, Portugal is seen as a hyperendemic region of human infection by Echinococcus granulosus, and hydatidosis is still of great epidemic and clinical significance in the Alentejo. Seeking to correct this situation, using the official statistics and our own case studies in the most endemic area of the country, we present a detailed analysis of the various parameters that will enable an updated assessment of the evolution of hydatidosis at national and regional levels.

When we began our investigations on hydatidosis in the district of Evora, we started with the obvious, i.e. on the definitive infection of the host, the dog (the only global study) is clearly outdated as it was conducted around forty years ago. With regard to animal hydatidosis, there was, until 1968, a large amount of official information on inspections in slaughterhouses, but nowadays, if such statistics exist, they are incomplete or inaccessible. In relation to human hydatidosis, the official data are clearly unsatisfactory; however, in conjunction with the case studies from Evora, the district with the highest incidence rates in the country, it is possible to build up a fairly accurate concept of the evolution of this parasitosis in Portugal, both temporally and spatially.

Key-words: hydatid disease, epidemiology, hydatid disease incidence, Portugal, Alentejo.
continuing previous studies, this paper consists of a longitudinal analysis of our own case studies.

Given that a significant number of National Health Service users undergo echography and other imaging exams, it can be said that nowadays, the professionals who tend to carry out the ‘screening’ for hydatidosis are Family Doctors – gone are the days when General Practitioners had no additional resources for diagnosis - therefore the cases diagnosed and referred to hospitals consist, in the majority of cases, of simple imaging findings, since the majority of hydatid cysts are asymptomatic, with only about 10% of patients actually being symptomatic.

**MATERIAL AND METHODS**

This study will analyze the statistical material from two different, but complementary sources:

a) Case studies attended by our Hydatidosis Consultancy, at Hospital do Espírito Santo de Evora (HESE): these comprise the medical records of 648 patients with hydatidosis, which we studied over a period of thirty years (1979-2008), including hydatidosis patients from the entire country.

b) official statistics: consisting of data available in the record of “Notifiable Diseases” and cover a period of twenty-one years (1987-2007) – 1987 being the year when notification of hydatidosis was made mandatory - and includes 467 hydatid patients.

In the analysis of the various parameters that we submitted to statistical analysis, we used, where appropriate, the $\chi^2$ test and calculations of confidence intervals (CI) at 95% – in order to avoid committing “(…) une erreur grossière: le résultat sous forme de pourcentage n’est pas accompagné de son intervalle de confiance. (…)” To determine the incidence of hydatidosis per 100,000 inhabitants per year, we used the official statistics for the period under study. When identifying the sites of infection, which is necessary, *verbi gratia*, to correctly determine the incidence per county, we used the “Military Map of Portugal”, with a scale of 1/25,000.

We wish to make our position clear with regard to the statistical analysis of our epidemiological case studies. In the official statistics – the “Notifiable Diseases” – doctors are required to declare, in the notification form, the patients’ place of residence. However, with the increasing migration of rural residents to urban and suburban areas (as in the case of the “Alentejo Diaspora”), many patients now live outside their place of origin where, in the majority of cases, they acquired the disease many years before. This epidemiological error is therefore reflected in the official statistics. Thus, when putting together the epidemiological history of each of our patients, we inquired, specifically, about the place of infection: the precise place where the patient was infected, if he/she had always lived in the place where they was born, or the probable place of infection, if the patient had lived in other places. We then identified, on the above mentioned “Military Map of Portugal” (the most detailed map available in the country), the place (hamlet, village or settlement) where the patient had acquired the infection, in order to assign the patient to the respective district or county.

Another point deserves mention: the number of cases registered in our consultation is higher than the number we have declared. Often we find patients with hydatidosis who, for instance, underwent surgery ten, fifteen or twenty years previously (often in Lisbon, to where the patients from the Alentejo used to be referred for surgery, or to where they had migrated for work), and who were now coming to see us due to a recurrence of the disease, or for clinical reassessment. Now, these cases have already been (or should have been) declared by the doctors who carried out the initial diagnosis, and therefore – a relevant fact – they do not represent new cases of hydatidosis. Thus, if we were to notify these cases now, after such a long period of time (usually decades) has elapsed since their diagnosis, this would surely contribute to an epidemiological error in the current statistics. Consequently, our “notifiable” cases of hydatidosis relate only to newly diagnosed cases.

**RESULTS**

**Country**

*Our case studies.* We shall analyze the most important parameters, from an epidemiological point of view:

a) The evolutionary pattern in the number of cases. Our cases studied over a 30-year period (1979-2008) include 648 cases of hydatidosis, of which 646 are national citizens and two are foreign citizens (from Turkey and South Africa). Since we are interested in characterizing the national epidemic-clinical scenario, we excluded from our analysis the two cases of hydatidosis acquired abroad, and will only statistically analyze the 646 clinical cases related to our coun-
and 1993. This fact was the result of the decision, by a director of our hospital, to refer patients with hydatidosis to our internists, whereas previously they were referred to our infirmary. This meant that many patients with hydatidosis were transferred to the Central Hospitals in Lisbon, or were not officially reported. However, in 1994, when we implemented the Consultation for Hydatidosis Cases, the situation returned to normal. To overcome this obstacle (which led to a drop in registered cases from 1990-1993), we analyzed the statistical data over five-year periods (Table I).

With the exclusion of that period of decreased notifications, it is observed that in Portugal, the notifications of hydatidosis cases increased in the late 1980s and early 1990s, then declined sharply: from 169 cases in the five-year period from 1984 to 1988, to 35 cases in the last five-year period, from 2004 to 2008.

b) Cases of hydatidosis by gender: 286 patients (44.3%; CI: 40.4-48.2) were male and 360 (55.7% – CI: 51.8-59.6) were female. The differences between genders were not statistically significant (p>0.05).

c) Cases of hydatidosis by age group. The patient distribution was as follows: Aged 0-9 years: 20 patients (3.1%); aged 10-19 years: 36 (5.6%); aged 20-29 years: 57 (8.8%); aged 30-39 years: 85 (13.2%); aged 40-49 years: 89 (13.8%); aged 50-59 years: 119 (18.4%); aged 60-69 years: 140 (21.7%); aged 70-79 years: 85 (13.2%); aged 80-89 years: 14 (2.2%); aged 90-99 years: 1 (0.2%). Fig. 2 shows the distribution of patients by age. The minimum and maximum ages were 2 and 96.

d) Possession of dogs: 447 patients (69.2% – CI:
65.5-72.7) reported current or previous possession of dogs in their home, or previous household, and 199 (30.8% – CI: 27.3-34.5) reported that they did not have dogs. The difference between both groups was statistically significant (p<0.05).

e) Cases of hydatidosis by sector of activity. The non-active population was relatively high: 315 patients (48.8%, CI: 44.8-52.7) – students: 7.7%; housewives: 13.5%; retired: 27.1%; and unemployed: 0.5%. The active population consisted of 331 patients, corresponding to 51.2% (CI: 47.3-55.2), with a slight majority in the primary sector (agriculture) (22.3%), closely followed by the tertiary sector (services) (21.5%), while the secondary sector (manufacturing) corresponded to only 7.4%.

f) Cases of hydatidosis according to the site of hydatid cysts. The analysis of this parameter has clear interest not only from a clinical point of view, but also from the epidemiological standpoint, since the infection of the two main organs (the liver in particular, and the lungs) is related to age distribution. The liver was the organ most frequently affected, in 85% of the cases, without any significant variations in relation to a previous study.6

Official case studies. In the report of “Notifiable Diseases”, we have access to statistical data from 1987-2007, i.e. a period of 21 years. During this period, 467 patients with hydatidosis were registered, which represents an average of 22 cases per year. The temporal evolution of the parasitic zoonosis is broadly represented in Fig. 1, since Evora is the district that has the highest number cases in the country. To overcome the inconvenience of the decrease in the number of cases notified from 1990 to 1993, the cases officially reported in Portugal should be grouped by five-year periods, which enables an increase in the cases of hydatidosis in our country in the 1980s and 1990s, followed by an irreversible decline, to be confirmed, as analyzed of Table I above shows.

Major Regions of the Country

Our case studies. Of the 646 cases of infection in Portugal, 565 (87.5% – CI: 84.7-89.9) were from the district of Evora and 81 (12.5% – CI: 10.1-15.3) from other districts of the country. The patients were referred to our Consultation for Hydatidosis by other doctors, or else the patients themselves took the initiative of seeking a consultation with an expert in hydatidology. The distribution by region of the 81 patients who did not acquire the infection in the district of Evora was, in ascending order of importance: Algarve, 3 patients (3.7%; all from the district of Faro); North, 4 patients (4.9%; 1 from Bragança and 3 from Vila Real); Central region, 5 patients (6.2%; 1 from Aveiro, 2 from Castelo Branco, 1 from Coimbra and 1 from Viseu); Lisbon and Tagus Valley, 10 patients (12.3%; 3 from Lisbon, 4 from Santarem and 3 from Setubal); and the Alentejo (excluding the district of Evora), 59 patients (72.8%; 25 from Beja and 34 from Portalegre).

Official case studies. Over the 21-year period for which official statistics are available, the number of cases increased in the major regions, as follows: Algarve 1.1%; North: 4.5%; Lisbon/Tagus Valley: 8.4%; Central Area: 9.2%; the Alentejo: 76.9%.

The Alentejo

Official case studies. In order to provide a better understanding of the importance of the parasitosis in the districts of the Alentejo and its evolution over the past 21 years, we compiled Fig.3 with the statistical data from the “Notifiable Diseases”. As mentioned earlier, the decrease in cases from 1990-1993 in the evolutionary curve for the district of Evora meant that the figures did not correspond to the epidemiological reality, but were the result of a failure to notify the disease, causing the number of cases of hydatidosis expected during this period to be misrepresented.

District of Evora

Incidence of hydatidosis in the three decades from
1979-2008. Table II shows the incidence of hydatidosis per 100,000 inhab./year in our case studies, calculating the annual average. The average incidence for the district corresponded to 10.7 cases/100,000 inhab./year. However, this figure deserves consideration: If in the general calculation we exclude, from the total population of the district (175,860 resident individuals) 42,769 people from the urban boroughs of the greater city of Evora (as stated in Material and Methods, for the thirty-year period we worked with the average figures given in the statistical Censuses for 1981, 1991, and 2001), the incidence rate for what we could call the “rural population” rises from 10.7 to 14.2.

Incidence of hydatidosis by five-year periods. In relation to the analysis of the evolution of the general incidence of hydatidosis in the district of Evora, it is clear that an analysis over five-year periods is necessary, since the incidence over the thirty-year ended up, necessarily, being marked by the high incidence in the 1980s and 1990s. Thus, as shown in Table III, the decline of cases of hydatidosis in the district, currently corresponding to rather modest values, is clear.

Incidence of hydatidosis by county. Table II shows the incidence rate of hydatidosis by county, calculating the annual average for the thirty year period covered by our case studies. For decreasing values, it was found that from 1979-2008, the county of Alandroal was the one with the highest number of (44.4 cases/100,000 inhab./year) while Vendas Novas was the county with the lowest number (0.9 cases). It is important to note that of the 53,984 inhabitants of the county of Evora – the largest urban county in the district – we excluded the 42,769 inhabitants of the urban boroughs. Thus, the overall incidence increases from 6.5 to 22.1 cases in the rural areas, so that this county, which was considered a mesoendemic region, now becomes hyperendemic.

Given the nature of dynamic transmission of hydatidosis over time, we decided to compare the inciden-

<table>
<thead>
<tr>
<th>Counties</th>
<th>Thirty-year period 1979-2008</th>
<th>Five-year period 2004-2008</th>
</tr>
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<tbody>
<tr>
<td></td>
<td>Inhabitants*</td>
<td>Hydatidosis cases</td>
</tr>
<tr>
<td>Alandroal</td>
<td>7 352</td>
<td>98</td>
</tr>
<tr>
<td>Mourão</td>
<td>3 330</td>
<td>24</td>
</tr>
<tr>
<td>Portel</td>
<td>7 647</td>
<td>38</td>
</tr>
<tr>
<td>Redondo</td>
<td>7 893</td>
<td>39</td>
</tr>
<tr>
<td>Arraiolos</td>
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<td>39</td>
</tr>
<tr>
<td>Borba</td>
<td>8 283</td>
<td>38</td>
</tr>
<tr>
<td>Vila Viçosa</td>
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</tr>
<tr>
<td>Reguengos de Monsaraz</td>
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<td>41</td>
</tr>
<tr>
<td>Viana do Alentejo</td>
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<td>19</td>
</tr>
<tr>
<td>Estremoz</td>
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<td>46</td>
</tr>
<tr>
<td>Évora</td>
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<td>105</td>
</tr>
<tr>
<td>Mora</td>
<td>6 477</td>
<td>12</td>
</tr>
<tr>
<td>Montemor-o-Novo</td>
<td>19 140</td>
<td>29</td>
</tr>
<tr>
<td>Vendas Novas</td>
<td>11 009</td>
<td>3</td>
</tr>
<tr>
<td>Total</td>
<td>175 860</td>
<td>565</td>
</tr>
</tbody>
</table>

ces occurring over the thirty-year period with more recent statistical data, i.e. data for the last five-year period (Table II), in order to understand the spatial and temporal evolution of endemic hydatidosis in the district. As for the thirty-year period, we also analyzed the incidence here, in parallel, excluding the urban boroughs of the city of Evora: thus, for the five-year period from 2004-2008, the overall incidence in the district rises from 3.2 (Table II) to 4.3 cases/100,000 inhab./year (which does not change the hypoendemic status of the district). In the county of Evora the incidences increases from 0.7 to 3.4 cases (which also does not alter its hypoendemic status).

**Cases of hydatidosis in the younger age groups.** Given the interest that the occurrence of hydatidosis in young patients assumes for the study of the evolution of the disease over time, we performed an analysis of cases diagnosed in young people aged 0-19 years, over five-year periods: 1979-1983: 10 cases; 1984-1988: 20 cases; 1989-1993: 14 cases; 1994-1998: 9 cases; 1999-2003: 3 cases; 2004-2008: 0 cases.

**Discussion**

The epidemiological field surveys are of great interest in developing or developed countries that have poor healthcare cover, particularly for rural populations. Therefore, when we began our work in the district of Evora in the 1970s, whether through hospital activity or the teaching in Human Parasitology as a discipline at the University of Evora, we began to conduct prospective parasitological field studies, through significant sampling of the resident population interested in hydatidosis and other parasitosis. During this time, the Portuguese rural population had access only to sporadic medical consultations at the “Casas do Povo”. Nevertheless, with the establishment of a National Health Services that was “universal and in general, tending to be free of charge”, virtually the entire population came under medical scrutiny, enabling the diagnosis of the more important pathologies. Methodologically, we leave prospective (cross-sectional) studies aside and conduct retrospective (longitudinal) studies, which provide a proper understanding of the evolution of a specific nosological entity. The present study was based on our clinical cases covering three decades — indeed, field surveys would never have provided us with a sample consisting of 650 cases of hydatidosis.

In the general context of infectious and parasitic diseases in our country (to which transhumance once clearly contributed to its dissemination), hydatidosis is the one for which the most detailed epidemiological information, both temporal and spatial, is available. The oldest study on hydatidosis that we were able to identify in various libraries around the country, dates back to 1864, a degree thesis. We then found an increasing number of publications, reflecting the attention that this parasitosis came to be given. As we showed in a previous work, from the early twentieth century, it became possible to measure the intensity of hydatidosis by analyzing the information on cases attended by at the Civil Hospitals of Lisbon, to which the vast majority of cases diagnosed in the country were referred. That study ended in 1978, but from 1979 on, we began our build up our own case studies, which allowed us to show, toge-

**TABLE III**

Hydatidosis cases (HESE): yearly average incidence in Evora district per five-year period

<table>
<thead>
<tr>
<th>Five-year period</th>
<th>Hydatidosis cases n</th>
<th>Average incidence / 100 000 inhab./year</th>
</tr>
</thead>
<tbody>
<tr>
<td>1979-1983</td>
<td>94</td>
<td>10.7</td>
</tr>
<tr>
<td>1984-1988</td>
<td>147</td>
<td>16.7</td>
</tr>
<tr>
<td>1989-1993</td>
<td>117</td>
<td>13.3</td>
</tr>
<tr>
<td>1994-1998</td>
<td>125</td>
<td>14.2</td>
</tr>
<tr>
<td>1999-2003</td>
<td>54</td>
<td>6.1</td>
</tr>
<tr>
<td>2004-2008</td>
<td>28</td>
<td>3.2</td>
</tr>
<tr>
<td>Total</td>
<td>565</td>
<td>10.7</td>
</tr>
</tbody>
</table>

**FIG. 3**

“Notifiable Diseases”: Hydatidosis cases in Alentejo, per district, per year.
ther with the official statistics, that cases of human hydatidosis was continuing to increase, reaching its highest rate of incidence in the late 1980s and early 1990s (Fig. 1). After that, we saw a gradual decline and stabilization of the number of cases in the last five-year period, with very modest values.

We shall now analyze the main evaluators of the evolution of human hydatidosis, and recall again that we used, simultaneously, two comprehensive sources of information, which reinforces the reliability of our analysis: the official statistics, covering a twenty-one-year period, and our own case studies, covering a thirty-year period.

**Total cases diagnosed**

**Country** Both our case studies (Table I and Fig. 1) and the official data are consistent with regard to the evolution of human hydatidosis in Portugal: following a growth in the rate of incidence since the beginning of last century, from the mid-1990s, this zoonosis declined sharply, and in the last five-year period, there have been, on average, only a dozen cases per year.

In the 1980s, we found a national incidence of 2.2 cases/100,000 inhab./year, which, broadly speaking, represents about a third that of our neighboring country for that period: “España mantiene la morbilidad de 6/100 000 H inconmovible desde 1985 (...)”. Thus, it is understandable that studies have been published in Spain involving case studies of, for example, 7435 patients with hepatic hydatidosis. Nevertheless, we also note some of the other incidence rates in Europe during the same period.

Corsica: 13 cases/100,000 inhab./year; Sardinia: 15 cases; Greece: 13.4 cases; Cyprus: 12.9 cases (before the eradication campaign), etc. It is also noted that in Africa, the Turkana region of Kenya, had an incidence of 220 cases (in fact, the real figure is probably even higher, since this figure is based only on cases actually operated on).

Taking the average rate for the last five-year period from the Portuguese official data (2003-2007), we find an incidence rate of 0.1 cases (52 cases in the mainland population, 9,869,343 patients – 2001 Census). Our country is, therefore, clearly hyperendemic. To become a hyperendemic region, these figures would have to be multiplied by a hundred! It is concluded that statistically, Portugal is not – and has never been - a hyperendemic country, neither was it mesoendemic (WHO defines as hyperendemic incidence rates ≥ 10 cases/100,000 inhab./year, and we define as mesoendemic and hypoendemic incidences as between 5-10 cases, and < 5 cases, respectively). Moreover, in addition to ≥ 10 human cases, WHO also considers as hyperendemic a region with incidences > 50% among sheep. However, in Portugal, over a quarter of a century (1944-1968) the percentage of rejected sheep viscera was only 2.2%! We hope, then, to definitively correct the stereotype, divulged internationally, that Portugal is or was a hyperendemic country.

**Major Regions of the Country.** According to the report of “Notifiable Diseases” (1987-2007), the Algarve was the region with the lowest incidence of hydatidosis (1.1% – CI: 0.3-2.5) and the Alentejo was the region with the highest incidence (76.9% – CI: 72.8-80.6) – North: 4.5%; Central region: 9.2%; Lisbon/Tagus Valley: 8.4%. If we correlate the average annual number of cases reported more recently (five-year period of 2003-2007 – “Notifiable Diseases”) for the respective population, we find the following incidence rates, in ascending order of importance: a) Northern Region: 0.02 cases/100,000 inhab./year (4 cases of hydatidosis in a population of 3,687,293 residents); b) Lisbon/Tagus Valley: 0.02 (3 cases in 2,661,850 people); c) Algarve: 0.05 (1 cases in 395,218 people); d) Central area: 0.1 (10 cases in 2,348,397 people); e) Alentejo: 0.9 (34 cases in 776,585 people).

It is therefore clear that for the official data, all the regions of the country are classified in the hypoendemic group, with extremely modest values indeed.

**The Alentejo.** Also according to official data, we found the following incidence per district in the Alentejo region in the last five-year period (2003-2007): a) Portalegre: 0.3/100,000 inhab./year (2 cases in 127,018 residents); b) Beja: 1.1 (9 cases in 161,211 residents); c) Evora: 2.6 (23 cases in 173,654 residents).

Thus, epidemiologically, all the districts of the Alentejo are currently classified in the hypoendemic group. At this point, we should also correct the fallacy that the Alentejo is a hyperendemic region. For example, from the “V Iberian Congress of Hydatidology”, held in Evora from November 5-7, 2008, the following information is published in the Congress website: “(...) This disease seems to have a higher prevalence in the Alentejo, including the counties of Elvas, Alandroal and Campo Maior, which have one of the highest prevalence rates of human hydatidosis in Europe.
(…)";25 and the original version of the site was even more assertive: “(…) This disease is hyperendemic in Alentejo, including the counties of Elvas, Alandroal and Campo Maior, which have one the highest prevalence rates of human hydatidosis worldwide. (…)”25 – emphasis added. However, the counties of Elvas and Campo Maior (about the county of Alandroal, see below) belong to the district of Portalegre – which also includes twelve other counties! –, where there were only two registered cases of hydatidosis in the last five-year period. Thus, the counties of Elvas and Campo Maior cannot, by any means, have “(…) one of the highest prevalence rates of human hydatidosis in Europe (…)”, much less “(…) one of the highest prevalence rates of human hydatidosis worldwide. (…)”. It is naturally, rather odd that the “Portuguese Society of Hydatidology” is the entity disseminating - or allo-carrying the dissemination through it – of misinformation on the epidemic situation in Portugal. The situation of hydatidosis in the district of Portalegre (Fig. 3) is therefore clarified: the district of Portalegre is mainly sine-endemic, with perhaps only two hypoendemic counties – Elvas and Campo Maior.

Regarding the county of Beja, it is sine-endemic in its western counties and hypoendemic (in general with rather low rates) in its eastern counties (Fig. 3).

**District of Evora.** For this district - the only on in Portugal that in clinical and epidemiological terms, can properly be called a former hyperendemic district2,9 - we will report data from our case studies, because they cover a wider timeline (thirty years), they contain a larger number of cases, and most importantly, they allow us to break down our epidemiological analysis into counties (in practice, our district monitoring is processed to borough level). This enables us to identify any active sources of parasitosis, and to study the individuals possibly involved in the infection, particularly family members, though this level is not of interest in our present analysis. Thus, in the thirty-year period 1979-2008 (Table II), the district would still be considered a hyperendemic region, with an average incidence rate of 10.7 cases (14.2 for the rural area – see above). Nevertheless, our analysis of the evolution of hydatidosis over time (Table III) showed that: in the five-year periods 1979-1983, 1984-1988, 1989-1993 and 1994-1998, the district was in fact hyperendemic; in the five-year period 1999-2003, it was mesoendemic; and in the last five-year period, it dropped to the category of hypoendemic. Breaking down our analysis, the statistical data shows that in the thirty-year period 1979-2008, 10 counties (Alandroal, Mourão, Evora rural, Portel, Redondo, Arraiolos, Borba, Vila Viçosa, Reguengos de Monsaraz and Viana do Alentejo) were still hyperendemic, three counties (Estremoz, Mora and Montemor-o-Novo) were mesoendemic and only one county (Vendas Novas) was hypoendemic. The cartographic representation of the isohydacid curves clearly showed the existence of a high concentration in the county of Alandroal, more specifically, in the suburb of Santiago Maior, with incidence rates gradually decreasing towards the west of the district.

However, as mentioned above, the incidence in the three-decade period was influenced by the great importance of the cases of hydatidosis in the 1980s and 1990s. Now, if we observe only the rates for the last five-year period (Table II), we see that: only the county of Alandroal had the characteristics of being considered hyperendemic; six counties (Redondo, Reguengos de Monsaraz, Mourão, Arraiolos, Borba and Estremoz) were mesoendemic; four counties (Vila Viçosa, Portel, Montemor-o-Novo and Evora) were hypoendemic; and no cases of hydatidosis were registered in three counties (Mora, Viana do Alentejo and Vendas Novas). Strictly speaking, we must assume that even the county of Alandroal, which was in fact hyperendemic, is no longer classified as such – in fact, as we have repeatedly shown, the high incidence rates in the county of Alandroal were due to the high incidence of hydatidosis in the suburb of Santiago Maior.2,9 In fact, the average age of the patients diagnosed with hydatidosis in the last five-year period in that county was 51 years; however, the average age for the 1979-1988 period, based on our case studies, was 43 years.2 Thus, the patients with hydatidosis considered new cases do not really represent new infections: the acquisition of the disease would have surely occurred some decades before, and now the transmission cycle of the zoonosis is very small, or has even stopped altogether, with only cases corresponding to old infections being diagnosed, but it is only now that studies on hydatidosis are being carried out (Fig. 2).

The evolution of hydatidosis over time, in the Alentejo, clearly has several causes: after the unsuccessful “Wheat Campaign” in the initial Salazarian period – “(...) Grâce aux ‘merveilles’ d’une ‘campagne
du blé’ (…) le pays est encore déficitaire en blé mais il est devenu ‘excédentaire’ en érosion (…). On a défriché des milliers et milliers d’hectares pour avoir, peu de temps après, la production de blé par hectare la plus basse de l’Europe (…) 26, –, powerful landlords moved towards extensive livestock farming, particularly sheep, but it followed traditional methods, i.e. with rural shepherds assisted by sheepdogs. Due to the lack of health education among the population, the “clandestine” slaughtering of sheep began to become a source of reinfection in dogs which, in turn, either by infecting the environment or by direct contact, infected human beings with eggs of E. granulosus. However, later, with the changes in the rural areas due to corporatization and globalization, several factors ultimately contributed to a drastic reduction in the transmission of the zoonosis in question, namely: a) in terms of employment, the primary sector, which was once almost exclusive, began to assume a residual position, b) the exodus of the Alentejo population to the outskirts of cities and even abroad was more pronounced (we have several patients who were operated on for hydatidosis in England, France and Germany); c) the education of young people caused them to move away from their traditional participation in the cattle farming; d) the aging of the population led to large numbers of workers retiring or going to live in retirement homes; e) wire fences were installed in cattle farms, relegating shepherds and sheepdogs to a secondary role; f) the improved sanitary conditions in the livestock sector, due in particular to the sale of meat products in supermarket chains, etc.

Overall age distribution
We often hear in conferences and/or read in articles and books, particularly those in the area of surgery, that hydatid cysts grow “1 cm per year” 27: for example, a hydatid cyst of 5 cm would supposedly be a result of an infection that occurred five years previously. This is another fallacy that needs to be deconstructed. Our long personal experience enabled us to observe cysts that, for two or three decades, had not grown (although they had undergone a process of organization – see next item). On the other hand, there were cases in which cysts grew considerably in size in just a few months. All this has to do with: a) the degree of viability of cysts, 28 b) the strain of the infecting E. granulosus, 2,29 c) the immunogenetic status of the host 30,31 (the hydatid cyst itself induces the production of blocking antibodies which slow down the growth of the existing cysts, so that the simple act of destroying a viable cyst may subsequently induce the growth of small cysts or oncospheres in dormant state – this is, in fact, one of the reasons justifying the prescription of therapy with benzimidazole after removal of the cyst), etc. We observed, then, that a hydatid cyst can grow 1 cm in a month, in a year, or in decades, or it can just stop growing at an unspecified time. It is important to remember that the medical literature often refers to a case of hydatidosis with a latency period of 53 years. 32 Nevertheless, we found two cases in which the latency period extended for at least 61 and 75 years, the time that had elapsed since these patients left Italy for the USA. 33,34

The above-mentioned fact lead us to the observation in Fig. 2, where we show that the group aged 60-69 years was the one with the highest number of cases of hydatidosis. In most countries, by contrast, cases of hydatidosis are observed long before this, in young and middle aged people. 12 In the group of patients aged 60-69 in our case studies, there are a significant percentage of retired individuals, who therefore are not exposed to the risk of infection. What actually happened was that those individuals would have acquired hydatidosis in their youth (from the family dogs or when, as it was common, they helped on the cattle farms) and then the cysts became dormant, only being diagnosed now, when family doctors are requesting abdominal ultrasounds more often.

Occurrence of hydatidosis in younger patients
There is a consensus that this is the most reliable parameter for the assessment of hydatid activity in a given region: if the infection remains active, then a significant number of cases occurs in children and adolescents; if the transmission cycle is less active or is eradicated, then there will be very few infected children, or no cases will be detected in the younger age groups. In the 0-19 year age group, we observed the following evolution over five-year periods: 1979-1983: 10 cases; 1984-1988: 20 cases; 1989-1993: 14 cases; 1994-1998: 9 cases; 1999-2003: 3 cases; and 2004-2008: 0 cases. The conclusion is clear: the zoonotic cycle of echinococcosis-hydatidosis is now very limited, or will have already been discontinued in most counties which were once considered endemic areas. In fact, something similar also occurred in the
second most endemic district of the country, Beja: “(...) The average age is high and no new cases have occurred in children in recent years. (…)”

We note that in areas of active transmission of the zoonosis, the distribution of cases of hydatidosis by age presents a bimodal distribution: for example, in 1056 hydatid patients from a hospital in Madrid in Spain, the age group with the highest number of infections was, notably, 0-9 years of age, followed by 40-49 years.

**Site of hydatid cysts by organ**

This point is related to the previous two. In effect, in the younger age groups, hydatid cysts are predominantly located in the lungs (there would be “immaturity” of the liver capillaries, the so-called “first physiological filter”, so the oncospheres would go to the “second physiological filter”, the lungs; or, according to some authors, there would be an alternative lymphatic derivation route), while in adults the cysts are predominantly located in the liver. However, our case studies clearly show the number of that cysts located in the lungs was reduced: 6.3%, versus 10.1% in the ten-year period of 1979-1988. And if we consider the most recent data, we recorded, for the last five-year period, only two cases of cysts in the lungs (5.7%) in all 35 cases of hydatidosis. For comparison, it is important to note the occurrence of pulmonary hydatidosis in areas of active transmission of the disease: of the 1056 cases recorded at a hospital in Spain, 24.5% had pulmonary hydatidosis, in Greece, of the 2000 cases, 30.3% of the cysts were located in the lungs.

**Evolutionary phases of hydatid cysts**

It is important to remember that the hydatid cysts are living entities; they are born, grow and die. So clinically, it is very important to assess their evolutionary phase, i.e., their viability, as the therapeutic options, are nowadays, dependent on this viability. Modern imaging techniques, in conjunction with immunology, facilitates the evaluation of hepatic cysts, which accounts for the majority of hydatid cysts. In fact, both the Gharbi classification and the WHO classification (which, after all, merely swapped the types II and III of the Gharbi classification) assess the evolutionary stages of the cysts, i.e., their degree of aging. In the two decades of our clinical study on hydatidosis, we did not observe any Gharbi type I cyst (CL, CE1 and CE2 of the WHO classification: “Active group: cysts developing and are usually fertile”). The cysts diagnosed were degenerating (Gharbi type II or WHO CE3: “Transition group: cysts starting to degenerate, but usually still contain viable protoscoleces”) or were mostly non-viable cysts (Gharbi types IV and V or CE4 and CE5 in the WHO classification: “Inactive group: degenerated or partially or totally calcified cysts – very unlikely to be fertile”), which is totally in keeping with the higher incidence of hydatidosis in older age groups and with older infections (Fig. 2).

**CONCLUSIONS**

Due to the lack of reliable and updated statistical information abroad, the totally false idea that Portugal is perhaps the last hyperendemic country in Europe is still widespread. Besides, within the country, there is a tendency to convey information that human hydatidosis in the Alentejo continues to be of great prevalence (we do not know the true motivations behind this statement). In terms of incidence, the vast majority of counties in Portugal are now epidemiologically classified as sine-endemic. In Portugal, the epidemiological scenario that is disseminated in scientific meetings on hydatidosis is indubitably an outdated one: methodologically, only the statistics for the last few years (see last five-year period) give an accurate picture of the current situation in the country, enabling us to outline a strategy for combating the zoonosis. Thus, the “need” for a campaign against echinococcosis-hydatidosis in the country, which we have seen being advocated in various forums, is not really necessary at all, when we consider the epidemiological data available: a simple cost-benefit analysis easily shows that it lacks any purpose whatsoever. Of course this does not eliminate the need for regular and effective campaigns to combat the parasitosis in dogs - the first links in the chain of contamination - which, essentially, should be accompanied by the essential prevalence studies, something that unfortunately is not normally done.

We conclude by offering a suggestion to the health authorities: in our case studies, we analyzed the results based on the place of infection, while the statistics in the “Notifiable Diseases” are based on the patients’ place of residence. However, while it is epidemiologically important to know whether a patient resides in avenue “x” or “y” of a given place, it is also (particularly) important to us, as epidemiologists, to
know where the source of infection of hydatidosis, swine flu, brucellosis, hepatitis, etc. may be. Thus, in our personal understanding, the “Notifiable Infectious Diseases” form should include, in addition to the obvious place of residence of the patient, the (exact or estimated) place of infection.

References

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